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Right atrial thrombus mimicking myxoma in antiphospholipid syndrome with secondary immune thrombocytopenic purpura

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Abstract

A 22-year-old woman was referred to our hospital due to a large right atrial mass. Echocardiography showed the mass to be pedunculated, mobile, partially calcified, internally heterogeneous, and with a rough surface. Computed tomography showed internal calcification and heterogeneity. On magnetic resonance imaging, the mass was isointense to hypointense on T1-weighted images and hypointense to hyperintense on T2-enhanced images. Simultaneously, laboratory work-up revealed antiphospholipid syndrome (APS) with secondary immune thrombocytopenic purpura (ITP). We could not definitively differentiate between myxoma or cardiac thrombus associated with APS. Preoperatively, the patient was administered γ-globulin and methylprednisolone due to ITP. Surgical resection of the mass was performed in order to avoid incarceration to the tricuspid valve or embolization. Based on gross and histologic findings, the cardiac mass was diagnosed as a thrombus. She was discharged with no postoperative bleeding. APS-associated cardiac thrombus is rare. It is difficult to distinguish APS-associated cardiac thrombus from myxoma. Moreover, ITP sometimes occurs as a complication in patients with APS. We experienced a case of right atrial thrombus mimicking myxoma in a patient with APS and secondary ITP.

Key words：Cardiac thrombus, Antiphospholipid syndrome, Immune thrombocytopenic purpura, Myxoma

Introduction

Cardiac myxoma is the most common primary tumor of the heart. If a cardiac tumor is pedunculated on imaging studies, we usually suspect the tumor to be a myxoma. Only a few cases of cardiac thrombus mimicking myxoma associated with antiphospholipid syndrome (APS) have been reported. It is difficult to distinguish cardiac thrombus from myxoma using imaging studies; however, the treatment strategies for each are different. In the case of APS-associated cardiac thrombus, anticoagulation is needed to prevent thrombus enlargement and embolization. Additionally, secondary immune thrombocytopenic purpura (ITP) sometimes occurs as a complication in patients with APS; thus, it is necessary to treat thrombocytopenia due to ITP to prevent bleeding in the perioperative period. Here, we report a case of APS-associated right atrial throm-

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bus mimicking myxoma with secondary ITP.

Case report

A 22-year-old woman was taken to a nearby hospital by ambulance due to a sudden onset of abdominal pain. When she was 20 years old, she was first diagnosed with thrombocytopenia, but the etiology had remained undetermined. She was followed up via observation because she was asymptomatic.

She underwent computed tomography (CT) at a nearby hospital. Although there was no abnormality of the abdominal organs, imaging revealed a calcified nodule on the posterior wall of the inferior vena cava (IVC) and a giant mass with calcification in the right atrium. Echocardiography showed normal cardiac contraction and no cardiac valvular disease; a giant, pedunculated, mobile, 40-mm mass was present in the right atrium. The mass showed a rough surface, internal heterogeneity, and partial calcification (Fig. 1). She was diagnosed with cardiac myxoma and referred to our hospital, due to the risks of tricuspid valve incarceration and embolization to the pulmonary artery. Her abdominal pain resolved without treatment after a few days.

We conducted further imaging studies. Enhanced CT showed that the mass extended from the posterior wall of the right atrium to the tricuspid valve. The contour was lobular and rough. Internal attenuation was lower than that of the myocardium, with heterogeneity and calcification (Fig. 2). The mass was not enhanced. There was embolization in the lower branch of the left pulmonary artery. On magnetic resonance imaging (MRI), the mass appeared to arise from the atrial septum. The mass was isointense to hypointense relative to myocardium on T1-weighted images (Fig. 3A), and hypointense to hyperintense on T2-weighted images (Fig. 3B). Ultrasound showed that there was no deep venous thrombosis.

Laboratory work-up revealed thrombocytopenia, with a platelet count of 3.1 x 10^4/μL (Table 1), and elevated platelet-associated IgG (Table 2). Bone marrow biopsy showed increased numbers of megakaryocytes and decreased platelet production. Additional laboratory tests revealed elevated anticardiolipin β2 glycoprotein I complex antibodies and lupus anticoagulant (Table 2). She was, therefore, diagnosed with antiphospholipid syndrome with secondary immune thrombocytopenic purpura.

Although we could not make a definitive diagnosis of the cardiac mass as a myxoma or APS-associated cardiac thrombus, we scheduled surgical resection of the mass to avoid incarceration to the tricuspid valve or embolization. Preoperatively, we administered an anticoagulant (dalteparin sodium, 50 IU/kg/day) because of the possibility of APS-associated cardiac thrombus, intravenous immunoglobulin therapy (2.5 g/day) on preoperative days 9 to 5, and steroid pulse therapy (methylprednisolone 250 mg/day) on preoperative days 2 and 1 to prevent surgical bleeding due to ITP.

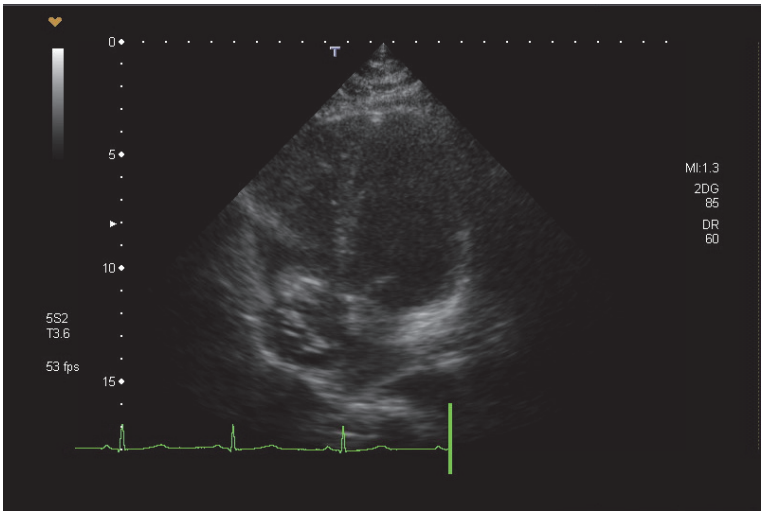

Fig. 1 Four Chamber View on Echocardiography

Echocardiography showing a giant, mobile, pedunculated, 40-mm mass, with a rough surface, internal heterogeneity, and partial calcification in the right atrium.

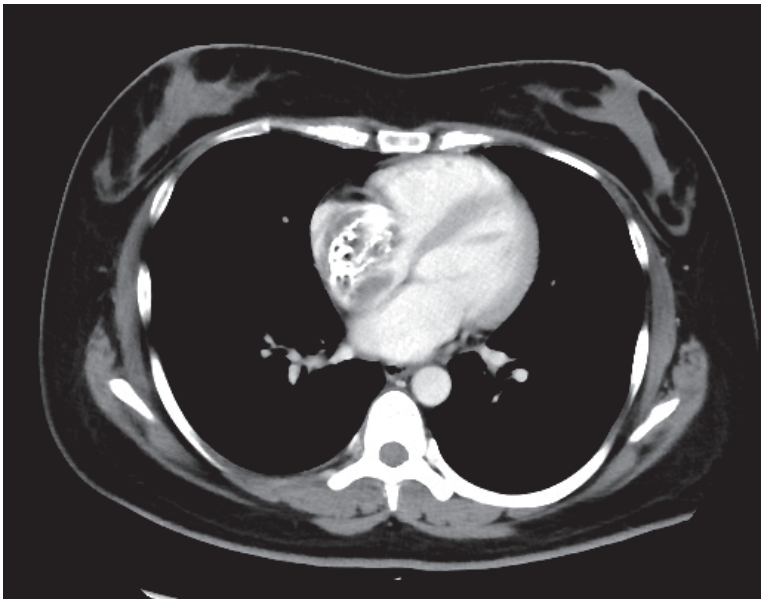

Fig. 2 Enhanced Computed Tomography

Enhanced computed tomography showing the mass extending from the posterior wall of the right atrium to the tricuspid valve. The contour was lobular and rough. The internal attenuation was lower than that of the myocardium, with heterogeneity and calcification.

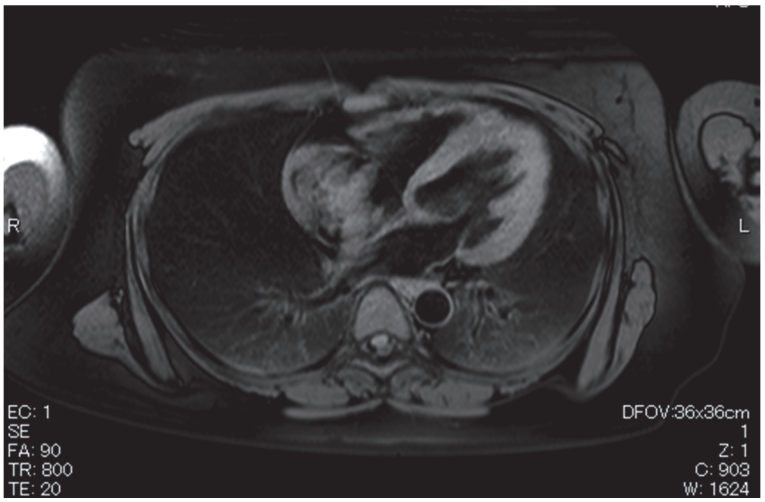

Fig. 3A T1-Weighted Magnetic Resonance Image

T1-weighted magnetic resonance image showing the mass to be isointense to hypointense relative to myocardium.

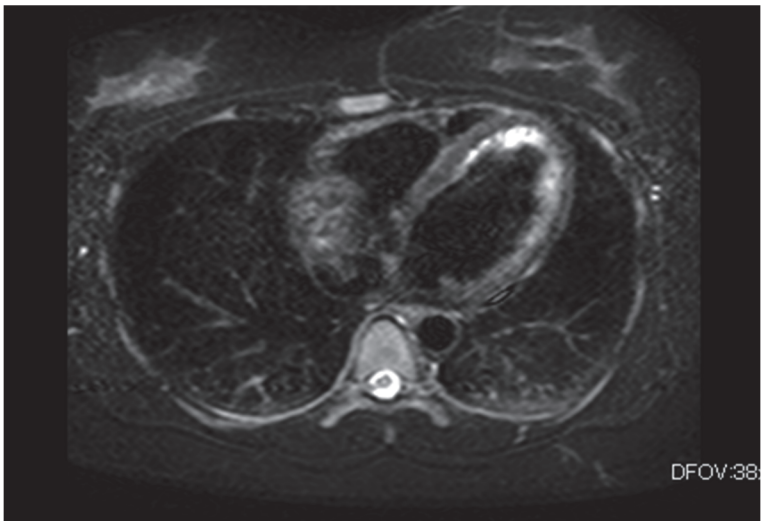

Fig. 3B T2-Weighted Magnetic Resonance Image

T2-weighted magnetic resonance image showing the mass to be hypointense to hyperintense relative to myocardium.

Table 1.

Complete blood count and coagulation tests	Results	
White blood cell count	$3.2x10^3$	/μL
Red blood cell	$4.03x10^6$	/μL
Hemoglobin	12.5	g/dL
Hematocrit	35.3	%
Platelet count	$3.1x10^4$	/μL
Prothrombin time (international normalized ratio)	0.99	
Activated partial thromboplastin time	47.4	Sec
Fibrinogen	194	mg/dL
Fibrin degradation products	5.1	μg/mL
Antithrombin III	98	%

Table 2.

Laboratory tests (autoimmune)	Results		Normal range
Soluble interleukin-2 receptor	410	U/mL	150-505
Platelet associated immunoglobulin G	491	ng/10^7cells	0-46
Anti-cardiolipin-beta2-glycoprotein I complex antibody	9.1	U/mL	0-3.5
Lupus anticoagulant	2.25		0-1.3
Comlement 3	73	mg/dL	73-138
Comlement 4	11	mg/dL	11-31
50% hemolytic complement activity	32.2	U/mL	24.2-52.8
Rheumatoid factor	≦7	U/mL	0-15
Anti-nuclear antibody, quantitative	40		≦40
Anti-DNA antibody	7.6	IU/mL	0-6
Anti-double stranded DNA immunoglobulin G	10	IU/mL	0-12
Anti-single stranded DNA immunoglobulin G	155	AU/mL	0-25
Anti-ribonucleoprotein antibody	negative		
Anti-Sm antibody	negative		
Anti-SS-A/Ro antibody	negative		
Anti-SS-B/La antibody	negative		

The surgery was performed via the median sternotomy approach under cardiopulmonary bypass (CPB). In order to avoid embolization of the cardiac mass, venous cannulae were inserted from the superior vena cava and right femoral vein. Under cardiac arrest, the right atrium was opened. The giant mass showed an irregular and fibrin-rich surface, and appeared grossly similar to an old thrombus (Fig. 4A, B). It was attached to the free wall and the septal wall. The mass was removed as much as possible to avoid leaving any residual tissue. A second small mass with a

smooth surface was attached to the posterior wall of the IVC and was also resected. Platelet transfusion was administered after weaning of CPB, and the operation was completed without severe bleeding.

Histologic examination showed the giant mass to be amorphous with no cellular component (Fig. 5), and partially replaced by collagenous fibrous tissue or calcified tissue. The small mass showed similar histologic findings, but collagenous fibrous tissue was more prominent. The small mass was considered older than the giant mass. From the gross and histologic findings, the cardiac mass was diagnosed as a thrombus.

We initiated warfarin therapy (international normalized ratio 2.0-3.0) beginning on postoperative day 1. Postoperative course was uneventful. She was discharged from the hospital 16 days after surgery and has had no relapse in the 2-year follow-up.

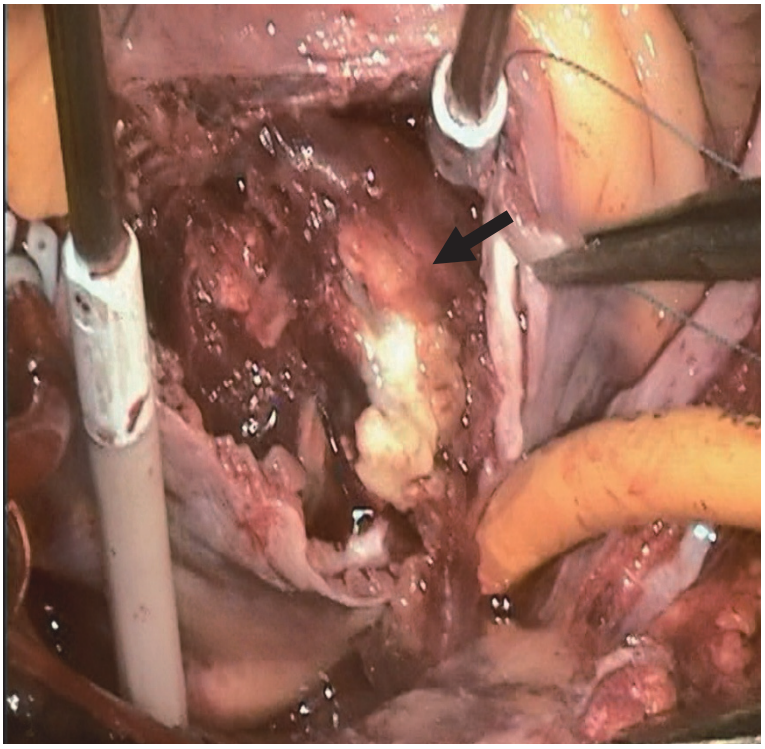

Fig. 4A Intraoperative View

Operative view showing the giant mass with irregular and fibrin-rich surface (arrow) in the right atrium.

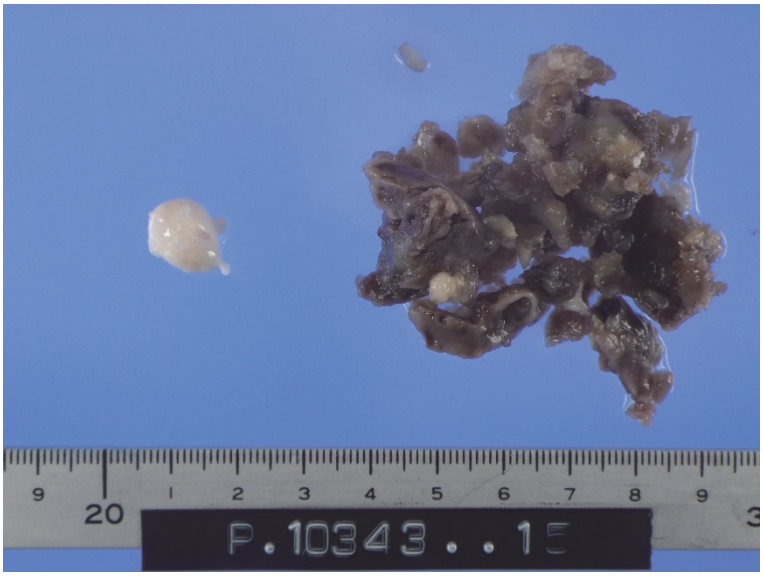

Fig. 4B Macroscopic Finding

Macroscopic finding. The small mass (left) with a smooth surface was attached to the posterior wall of the inferior vena cava. The giant mass (right) was in the right atrium.

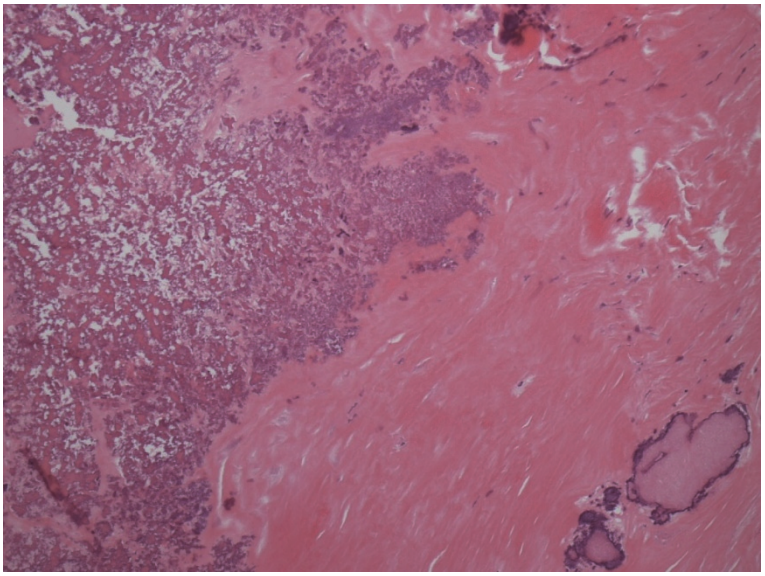

Fig. 5 Pathological Tissue

Histopathological examination showing the mass to be amorphous and with no cellular component and was partially replaced by collagenous fibrous tissue or calcified tissue. Picture was taken through a ×40 objective.

Discussion

Myxomas are the most common tumors of the heart, representing approximately 40% to 83% of all primary cardiac tumors [1,2]. Pathologically, myxomas are of endocardial origin; their cells are considered to be multipotential mesenchymal cells that persist as embryonal residues during the septation of the heart. Sixty percent of myxomas originate in the left atrium, and 28% in the right atrium [3]. Most myxomas are pedunculated and mobile, and arise from the atrial septum at the border of the fossa ovalis. The appearance is most commonly round or oval, with a smooth or lobulated surface. If a cardiac tumor has a pedicle and is mobile, the suspected diagnosis is cardiac myxoma. Our patient was initially diagnosed with cardiac myxoma at a nearby hospital.

On the contrary, patients with APS rarely develop cardiac thrombus. APS is a systemic autoimmune disease characterized by arterial and venous thrombosis and/or recurrent fetal loss in the presence of lupus anticoagulants and/or anticardiolipin antibodies. APS affects the cardiovascular system. In a series of 1000 patients with APS, cardiac manifestations were observed in 27% of patients, mostly commonly characterized by valvular (14%) and coronary artery disease (8%) [4]. However, APS-associated cardiac thrombus is rare. Some reported cases of APS-associated cardiac thrombus developed in the right atrium [5-7] and left ventricle [8,9]. Pliakos and coworkers also reported right atrial myxoma coexisting with APS [10]. Thus, it is difficult to differentiate between myxoma and thrombus if the patient has APS. In particular, when the mass is in the right atrium, as in this case, it tends to be diagnosed as myxoma initially and determined to be thrombus after surgery [6,7]. Preoperative diagnosis is difficult due to similar appearances on echocardiography.

On echocardiography, the attachment site of the mass is useful in the differentiation of myxoma from thrombus. Myxomas almost arise from the atrial septum; in contrast, APS-associate thrombi can be adherent to the atrial septum, free wall, tricuspid valve and/or IVC.

MRI findings are also useful in the differentiation of myxoma from thrombus. Grebenc and coworkers reported the MRI findings of myxoma [11]. Myxomas showed heterogeneous signal intensity and were predominantly isointense to hyperintense with respect to the myocardium on T1-weighted images. T2-weighted images demonstrated heterogeneous and hyperintense signals. In this case, the cardiac thrombus differed in that it showed hypointense to isointense signal on T1-weighted images and hypointense to hyperintense signal on T2 images.

With regard to treatment strategy, myxomas should be resected as soon as possible to avoid embolization; if the myxoma arise from the septum, the atrial septum should be also resected to prevent recurrence. In contrast, in patients with APS-associated thrombi, it remains a matter of debate whether administration of anticoagulants

or surgical excision is the better approach. Administration of anticoagulants is necessary to prevent further thrombus expansion. However, in the case of incarceration of a thrombus to the tricuspid valve or giant thrombus, there is likely to be a significant risk of pulmonary embolization, and the thrombus should be surgically resected. APS-associated thrombus may attach to multiple intracardiac sites, as in the current case. Attention must be paid to ensure there is no residual thrombus. Postoperatively, echocardiographic follow up is necessary for both myxoma and APS-associated thrombus, because both may recur.

Thrombocytopenia is frequently observed in patients with APS, with a reported prevalence of 22% to 42% [12]. The pathophysiology of APS-associated thrombocytopenia is considered to be a similar to that of ITP. Our patient had severe thrombocytopenia. To prevent intraoperative bleeding, intravenous immunoglobulin therapy, splenectomy, steroid therapy, and platelet transfusion are often performed [13]. Our patient was administered γ-globulin at a dose of 2.5 g/day for 5 days preoperatively, methylprednisolone at a dose of 250 mg/day for 2 days preoperatively, and platelet transfusion after weaning of CPB. Her platelet count was maintained over 10 x 10^4/μL after surgery until discharge, and bleeding was well controlled.

Conclusion

We experienced a rare case of intracardiac thrombus associated with APS. It was difficult to preoperatively distinguish thrombus from myxoma. Secondary ITP with APS was also observed; and ITP treatment (intravenous immunoglobulin therapy, steroid pulse therapy and platelet transfusion) was necessary to prevent intraoperative bleeding. Following thrombus resection, an anticoagulant agent was administered, and she has had no relapse in the 2-year follow-up.

Conflicts of interest

All authors have no conflicts of interest.

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