



## Case Report

## Thrombocytopenia recovered by surgical resection of a myxoma

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## ABSTRACT

Myxoma is the most common primary cardiac neoplasm, and causes a variety of symptoms, including hematological disorder. An 82-year-old man with anorexia was diagnosed with a gastrointestinal stromal tumor. Computed tomography and echocardiography showed a 2-cm tumor in the left atrium. The patient had a history of lung and skin sarcoidosis, and interstitial pneumonia. Laboratory examination showed thrombocytopenia with a platelet count of  $23 \times 10^3/\mu\text{L}$  and elevation of IgA and platelet-associated IgG (PAIgG). We suspected that the thrombocytopenia was caused by the left atrial tumor. He successfully underwent resection of the tumor with cardiopulmonary bypass. The platelet count increased to  $166 \times 10^3/\mu\text{L}$  after surgery. Pathological examination showed Alcian blue staining of the extracellular and intracellular matrix, suggesting a mucopolysaccharide matrix. Immunohistochemical examination of the tumor revealed expression of CD31, CD34, and calretinin, which was consistent with a myxoma. The PAlG level decreased to the normal range at 36 days postoperatively. Thrombocytopenia is a relatively rare finding in patients with myxomas. However, in addition to mobility of the mass, thrombocytopenia should be kept in mind as an indication for surgery.

**<Learning objectives:** Patients with myxomas have various symptoms and abnormal laboratory values. Thrombocytopenia was seen in an octogenarian patient diagnosed with a myxoma. After surgical resection of the tumor, the platelet counts increased to the normal range and the platelet-associated IgG level, which had been elevated before surgery, decreased to the normal range. The pathological findings were consistent with a myxoma. Thrombocytopenia should be kept in mind for surgical removal of myxomas.>

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## Introduction

Myxoma is the most common cardiac neoplasm. These tumors range in size from 1 to 15 cm in diameter. Approximately 80% of myxomas originate in the left atrium. Patients with myxomas develop cardiovascular symptoms (67%), systemic embolization (29%), and constitutional symptoms (34%) [1]. Constitutional symptoms include fever (20%), anorexia, and unexplained weight loss (18%). Thirty-seven percent of patients with myxomas show laboratory abnormalities such as anemia, and a high erythrocyte sedimentation rate, C-reactive protein level, and/or globulin level [2,3]. About 10% of patients were incidentally diagnosed as occult tumor [1]. Although some cases of thrombocytopenia with myxoma have been reported, thrombocytopenia is a rare symptom

of myxoma. Resection is required because of the risk of embolization or cardiovascular complications. Some studies have reported an operative mortality rate of <5% [2,3].

## Case report

An 82-year-old man with a left atrial tumor was referred to our hospital for surgical removal of the tumor. Because he complained of anorexia at the first visit, he underwent esophagogastroduodenoscopy and was diagnosed with a gastrointestinal stromal tumor (GIST). He had history of lung and skin sarcoidosis, and interstitial pneumonia, all of which had been resolved. Computed tomography as a preoperative work-up examination before surgical removal of the GIST revealed a left atrial tumor. Echocardiography showed a 2-cm mobile tumor in the left atrium. The neck of the tumor was located on the atrial septum, and the morphology of the tumor was consistent with a myxoma. Laboratory examination showed thrombocytopenia (platelet count of  $23 \times 10^3/\mu\text{L}$ ) and slight elevation of IgA and platelet-associated

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IgG (PAIgG). The patient consulted with a hematologist and was diagnosed as having mild type of myelodysplastic syndrome (MDS). The patient had no signs of petechiae, mucosal bleeding, infection, thrombosis, or bleeding. His only medication was an antihypertensive drug. He had been previously healthy despite his age. We suspected that thrombocytopenia was mainly caused by the myxoma, because the platelet count was too low, considering severity of MDS. Therefore, we decided to surgically resect the tumor to prevent tumor embolism, and improve the thrombocytopenia.

Under general anesthesia, cardiopulmonary bypass was established through a standard median sternotomy. Because the tumor was fixed to the atrial septum, we assumed that a right side approach through a direct incision to the left atrium was unsuitable due to poor visibility. Therefore, we entered the left atrium through the right atrium by incising the fossa ovalis. We identified the tumor and extended the incision while avoiding the neck of the tumor. We removed the whole tumor with the inner membrane (Fig. 1). Twenty units of the platelets (Irradiated Platelet Concentrate, Leukocytes Reduced, NISSEKI, Tokyo, Japan) were transfused during surgery to control bleeding. The patient's postoperative course was uneventful and he was discharged on postoperative day 6. The platelet count increased to  $166 \times 10^3/\mu\text{L}$  before discharge without any additional transfusion. His platelet count decreased slightly after discharge. The platelet count was maintained above  $62 \times 10^3/\mu\text{L}$  for 5 months after removal of the tumor. The latest platelet counts was  $73 \times 10^3/\mu\text{L}$  (Fig. 2). Postoperative echocardiography revealed no tumor remnant or recurrence.

Macroscopic and microscopic examination of the tumor mass revealed bleeding, thrombosis and hemosiderosis (Fig. 3a,b). Pathological examination also showed Alcian blue staining of the extracellular and intracellular matrix, suggesting the matrix comprised mucopolysaccharides (Fig. 3c). An immunohistochemical study to exclude the possibility of angiosarcoma revealed expression of CD31, CD34, and calretinin (Fig. 3d). Pathologically, the cardiac mass was consistent with a myxoma. The PAIgG level decreased to the normal range 36 days after the operation (Fig. 2).

## Discussion

Myxoma is the most common primary cardiac neoplasm. Symptoms of myxomas are related to the cardiovascular system,

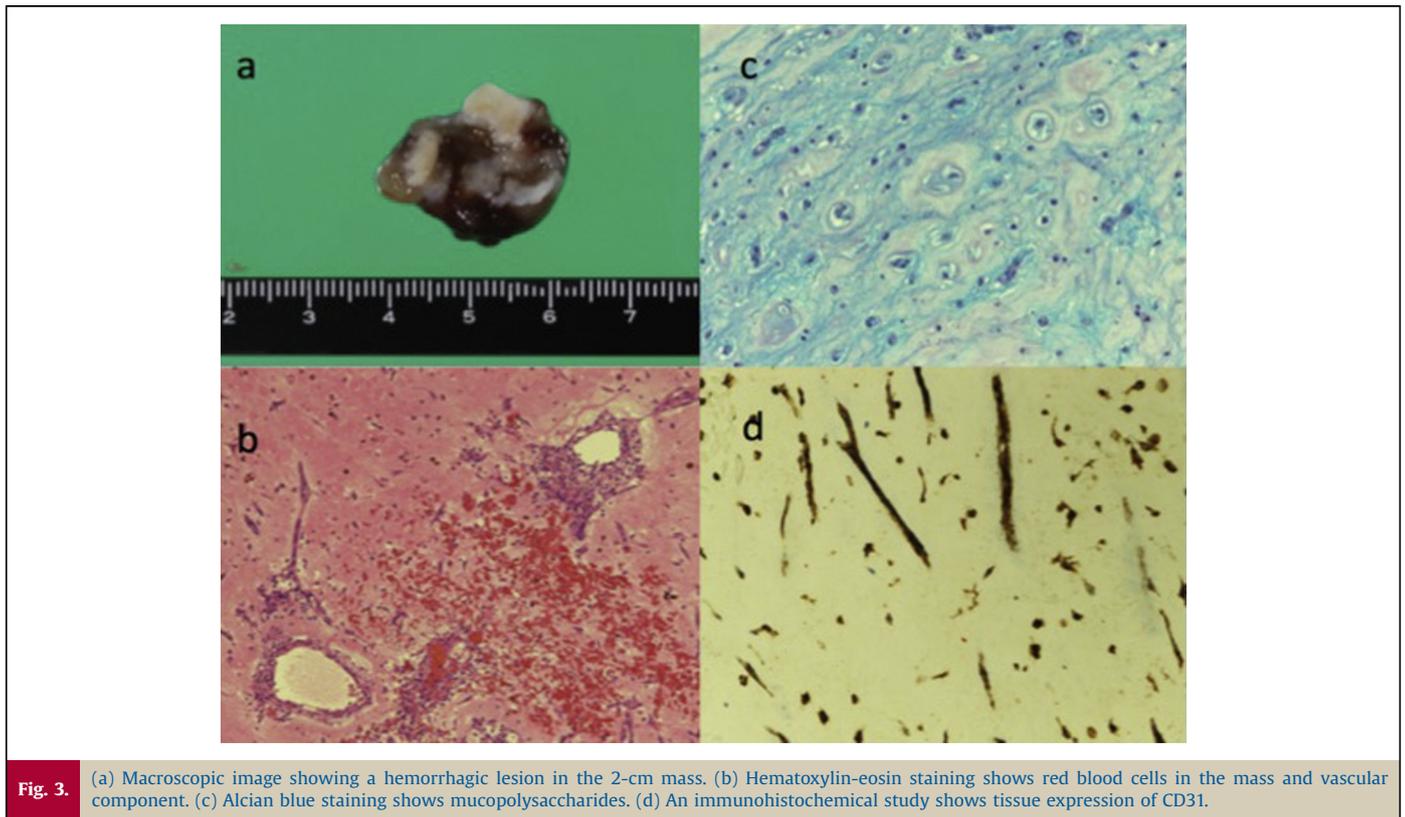
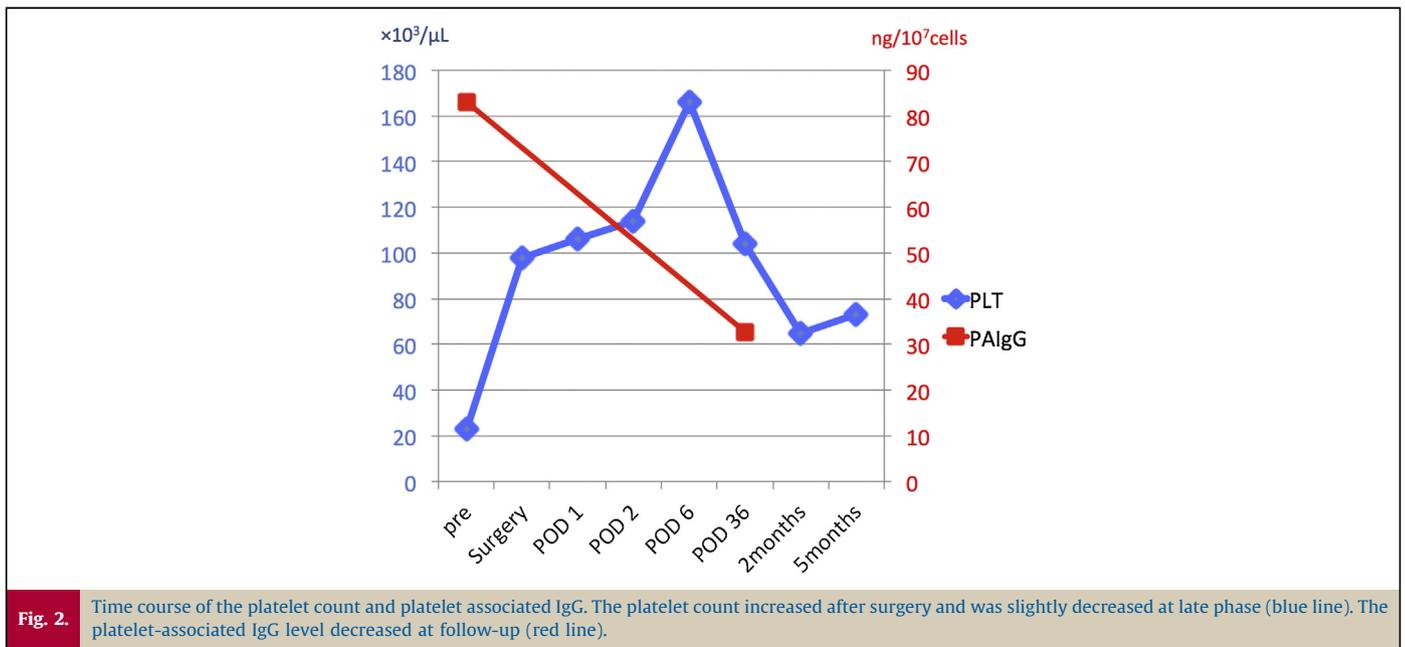
and systemic embolization and may also include constitutional symptoms (e.g. fever, weight loss). However, thrombocytopenia is less frequently seen. Because myxoma is associated with a risk of embolization or cardiovascular symptoms, including sudden death, surgical resection is the first-choice treatment. The outcome of surgical resection is satisfactory. In some studies, the operative mortality was less than 5% [2,3].

Thrombocytopenia is defined as a platelet count of  $<150 \times 10^3/\mu\text{L}$  and is caused by a variety of conditions. The platelet counts decrease by trapping of platelets in the spleen, or either decreased production or increased destruction of platelets. Thrombocytopenia confers a greater risk of bleeding. Severe thrombocytopenia is associated with a mortality risk of 4.2% [4,5]. In the present case, the patient's spleen was normal in size, and he had no history of heavy alcohol consumption, chemotherapy, or signs of infection. PAIgG, which was elevated in this case, is partially correlated with idiopathic thrombocytopenia (ITP) [6,7]. Therefore, the remaining explainable causes of the patient's thrombocytopenia were myxoma and ITP. The PAIgG level decreased to the normal range after the operation. If ITP had been the cause of the thrombocytopenia, the PAIgG would not have improved after the resection. Additionally, the patient's platelet count increased soon after surgery and remained within the normal range after resection of the myxoma. The patient's clinical course indicated that the myxoma was the main cause of the thrombocytopenia, although the MDS might have partly influenced the platelet count in this case. Platelet transfusion is another potential cause of a platelet count elevation. However, considering the average life span of circulating platelets is only 10 days [8], we determined that the patient's blood transfusion did not affect the elevation of his platelet count after the operation.

Thrombocytopenia can be associated with cardiac masses. Myxomas can cause thrombocytopenia and disseminated intravascular coagulation [9]. The mechanism of thrombocytopenia in patients with myxomas is unclear. An immunohistochemical study (CD31, CD34, and calretinin) was performed to exclude angiosarcoma in this case. The patient's myxoma expressed CD31, also known as platelet endothelial cell adhesion molecule-1, which is known to influence change in platelets. Additionally, the myxoma exhibited microscopic bleeding, thrombosis, and hemosiderosis (Fig. 3b), which might consume platelets. Therefore, the myxoma was the most probable explanation for the thrombocytopenia in this case.



Fig. 1. (a) Intraoperative picture through the fossa ovalis. The mass was attached to the atrial septum. (b) The tumor was round, and its surface was smooth. The mass was resected with the atrial septum, and macroscopic examination was consistent with a myxoma.



Surgical removal of myxoma is mainly indicated to prevent embolism formation and cardiac dysfunction. Echocardiography is important for risk assessment by evaluating the mobilization of the tumor and its potential influence on cardiac function. In the present case, in addition to a risk of embolism formation, we decided to resect it to improve the thrombocytopenia. Thrombocytopenia is associated with a risk of life-threatening hemorrhage. Surgical resection is a safe treatment as described above [2,3]. Considering the risk of hemorrhage and surgery, thrombocytopenia might be a reasonable indication for surgery, even in tumors with a low risk of embolism.

In conclusion, we have herein presented a case of resection of a myxoma in a patient with thrombocytopenia. The platelet count increased after resection of the myxoma. In addition to mobility of the mass, thrombocytopenia should be kept in mind as an indication for surgery.

#### Conflict of interest

We received no financial support and none of the authors had any conflicts of interest in regard to this report.

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