

**Case
Report**

Left Atrial Myxoma Complicated with an Acute Upper Extremity Embolism

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Atrial myxoma is the most common benign tumor of the heart. Patients who have atrial myxoma usually present with cardiac obstruction, arrhythmia, or peripheral embolization. We encountered an unusual case of acute upper extremity ischemia due to a massive atrial myxoma in a young man. A 38-year-old man was admitted to our hospital with an acute onset of severe, right upper extremity pain and paralysis while working. Neurologic examination yielded normal results, but the patient showed no palpable right radial or ulnar artery. Routine sonographic evaluation revealed acute aortic embolism in his right brachial artery. Because of his young age and otherwise healthy condition, we decided to perform trans-thoracic echocardiography, which showed a huge left atrial tumor, which we suspected to be myxoma. We then performed urgent concurrent open heart surgery and embolectomy to avoid further embolism. The microscopic findings of the resected tumor and embolism specimens were myxoma. He was discharged without complications.

Keywords: left atrial myxoma, acute aortic embolism, surgical resection

Introduction

Cardiac myxomas represent approximately 50% of all cardiac tumors, occurring mainly in the 3rd–6th decades of life. Of these, 85% are located in the left atrium (LA), with 15% in the right atrium (RA).¹⁾ Myxoma originates from the subendocardial mesenchymal cells, mainly in the LA. Patients with left atrial myxoma usually present with signs of cardiac failure. In some cases, it leads to loss of consciousness, signs of systemic embolism, or sudden death.

The embolization by tumor fragments or thrombotic material covered with tumor cells occurs in 30%–45% of

myxoma patients.²⁾ We report an unusual case of a young man who presented with acute upper extremity ischemia due to left atrial myxoma.

Case Report

A 38-year-old man had a medical examination because of chief symptoms of acute onset, right upper extremity pain and paralysis, while working. About 30 hours after onset, he was transferred to our hospital. His height was 175 cm, weight, 54 kg, and body temperature, 36.2°C. He had a resting pulse rate of 104 beats/min, and his blood pressure was 134/78 mmHg. He had no palpable right radial or ulnar artery, but his neurologic examination yielded normal results. Cardiac examination revealed sinus rhythm. No murmurs, rubs, or gallops were audible on auscultation. Electrocardiography showed sinus tachycardia and some supraventricular extrasystoles. Chest radiographic findings were normal. Laboratory findings showed a white blood cell count of 10.700/mm³ and a C-reactive protein level of 2.3 mg/dl. All other laboratory findings were normal. However, routine sonography showed an embolus in his right brachial artery just before

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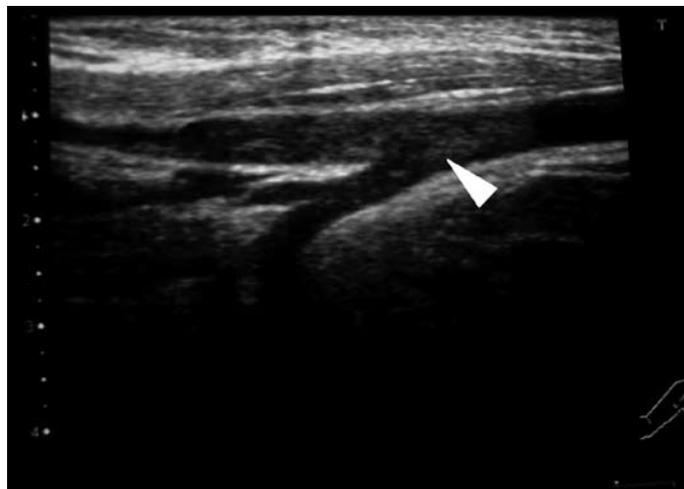


Fig. 1 Routine sonography showing an embolus in the brachial artery just before its bifurcation into the radial and ulnar arteries (arrowhead).

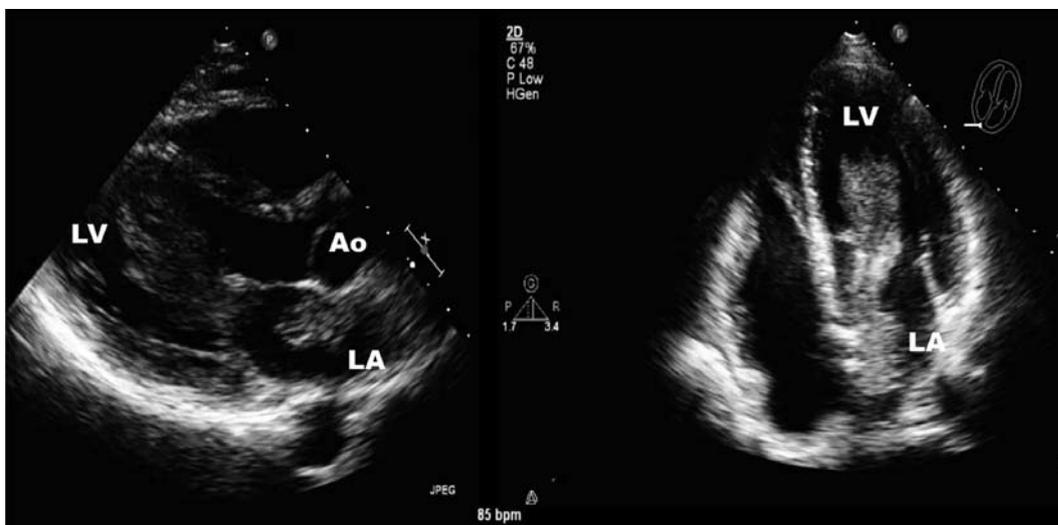


Fig. 2 Transthoracic echocardiography showing a huge tumor adhering to the interatrial septum. **a:** longitudinal axis view, **b:** 4-chamber view
LV: left ventricle; LA: left atrium; Ao: aorta

its bifurcation into the radial and ulnar arteries (**Fig. 1**). He was otherwise healthy, but he had palpitation on effort, 3 months previously.

We diagnosed acute arterial embolization, but we also performed transthoracic echocardiography (TTE) to exclude the possibility of cardiac embolism. TTE demonstrated a large $2 \times 3 \times 9$ -cm mass in the LA, which prolapsed through the mitral valve into the left ventricle (LV) in the diastolic phase, suggesting the diagnosis of atrial myxoma (**Fig. 2a** and **2b**).

We deduced that the cause of the acute arterial embolization was a cardiac tumor in the LA. To avoid further

neurologic injury and delay in treatment, preprocedural imaging was not performed. We performed emergency open heart surgery via a tumor resection. An embolectomy was performed through the right brachial and radial arteries using Fogarty catheters. The operation time was 205 min; the CPB time, 106 min; and the aortic cross-clamping time, 78 min.

The tumor was oval with a gelatinous, but non friable surface. It was $100 \times 28 \times 15$ mm in size and 110 g in weight. No thrombus was found on the surface, but the tip of the mass was partly torn (**Fig. 3a**). The embolus, which we removed from his brachial artery, was $30 \times 5 \times$

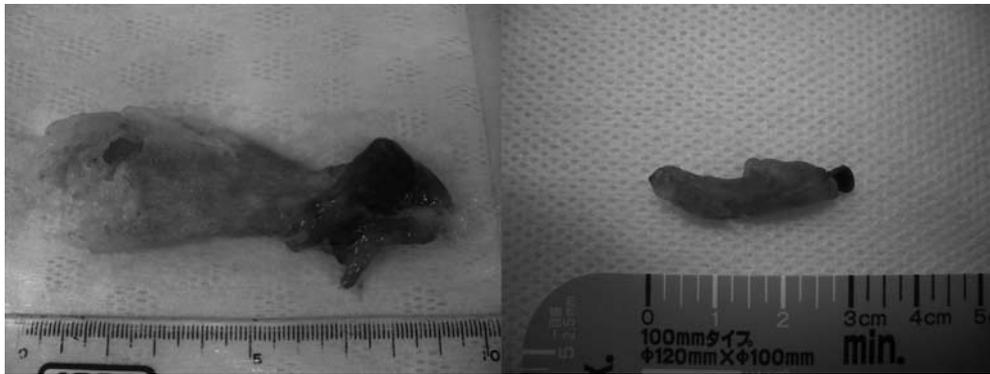


Fig. 3 Macroscopic appearance of the resected tumor (a) and extracted embolus (b), which shows an oval-shaped and gelatinous tumor.

a | b

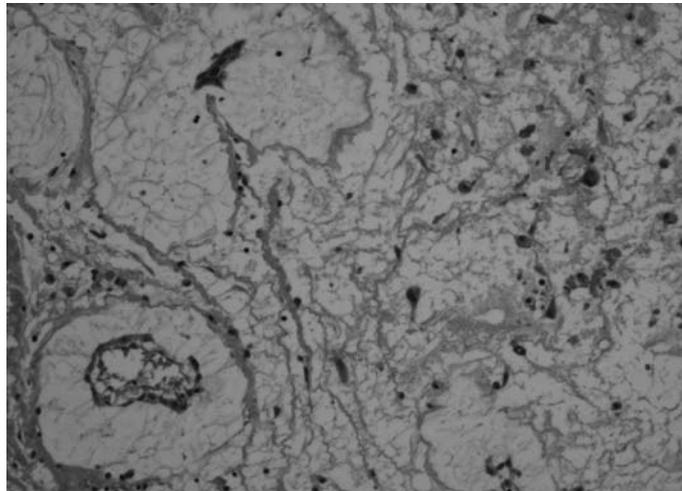


Fig. 4 Microscopic findings showing prominent spindle/ovoid/stellate cells around blood vessels. Hemosiderin pigmentation can also be seen.

4 mm in size, with a similarly sized collar (**Fig. 3b**).

Microscopically, prominent spindle/ovoid/stellate cells could be seen around the blood vessels, with a background of blue-gray mucopolysaccharide ground material (**Fig. 4**). Therefore, we diagnosed the tumor as a myxoma. The embolus, which we removed from the brachial artery, had identical microscopic findings.

After surgery, the patient was discharged from hospital without complications. One year after the initial event, he was in good health, without recurrence of cardiac myxoma.

Discussion

Atrial myxoma is the most common, benign tumor of the heart, followed by lipoma and papillary fibroelastoma. Clinical manifestations of atrial myxomas are de-

termined by their location, size, and mobility. Intracardiac obstruction and constitutional symptoms - fatigue, arrhythmia, erythematous rash, fever, arthralgia, and weight loss - are the most reported abnormalities.¹⁻⁵ A case of left atrial myxoma, complicated with acute upper extremity ischemia, such as in the present case, is very rare, and to the best of our knowledge only 10 cases have been reported in the literature among 4396 reports of myxoma.⁶

Embolization resulting from fragmentation or complete tumor detachment occurs in up to 30% of cases.¹ Most of these tumors embolize the central nervous system, resulting in cerebrovascular accidents; secondary embolization of coronary arteries, kidneys, intestines, and peripheral arteries also occur.⁷ In the present case, we did not scrutinize the patient's whole body computed tomography findings because the results of a physical

examination were normal except for his right upper extremity ischemia, and there were no laboratory findings which indicated an ischemic organization.

In cases such as the present one, the diagnostic and surveillance method of choice is echocardiography. Magnetic resonance imaging (MRI) is also useful in specific circumstances to distinguish cardiac tumors and provide additional information about the extent of tumor involvement.⁸⁾ Myxoma originates from subendocardial mesenchymal cells and is heterogeneous, and MRI shows hypointensity of the mass relative to the myocardium on T1-weighted images, and hyperintensity on T2-weighted images.¹⁾

Coronary angiography (CA) may also be useful in the further differentiation between cardiac tumors and thrombi.¹⁾ Parwis et al. suggested that CA should only be performed in patients over 40 years old, particularly those with atherosclerotic risk factors who require surgical removal of a cardiac mass.¹⁾

Although rare, cardiac myxoma may recur after surgical resection and is estimated to be less than 3% in solitary tumors and 12% to 22% in familial forms.⁷⁾ Therefore, echocardiographic follow-up within 1 year is very important. Familial myxoma can occur as part of the Carney complex, an autosomal-dominant inherited syndrome characterized by spotty skin pigmentation and endocrine tumors (especially adrenocortical disease, which is responsible for Cushing syndrome).⁷⁾ The current patient had no familial or personal history of endocrine disease.

In the present case, acute, upper extremity ischemia was due to the huge left atrial tumor, which was confirmed by microscopic findings. An embolic myxoma should be included in the differential diagnosis in young and otherwise healthy patients who present with acute arterial ischemia,⁶⁾ as in the present case.

Conclusion

We reported a rare case of acute, upper extremity ischemia in a 38-year-old man due to an embolic episode of a

huge left atrial myxoma, which we treated by embolectomy and open heart surgery.

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Disclosure Statement

The authors declare no conflicts of interest associated with this study.

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