

# Intimal Sarcoma: An Extremely Rare Case of a Left Atrial Tumor with Partial Obstruction of the Mitral Orifice



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## INTRODUCTION

Approximately 70% of left atrial tumors are identified as myxomas.<sup>1</sup> Myxomas exhibit a variety of echocardiographic features, such as their globular pedunculated nature with insertion point at the interatrial septum. They can also be quite large, resulting in obstruction of the atrioventricular valve. When a large left atrial myxoma prolapses into the mitral valve orifice during diastole, the mass can deform.<sup>2</sup> Although myxoma are benign in nature, approximately 25% of primary cardiac tumors are malignant. Echocardiography is the most convenient noninvasive imaging modality, but evaluation of the benign or malignant nature of detected masses is difficult. Few echocardiographic features of malignant primary cardiac tumors have been previously reported, and these have not yielded clarity. Here, we report a case of a large left atrial tumor mimicking a myxoma with partial obstruction of the mitral orifice.

## CASE PRESENTATION

A 45-year-old man presented with cough and palpitations for approximately 10 months before referral. The symptoms did not diminish and were accompanied by weight loss and hyperhidrosis; therefore, he was referred to a secondary hospital by his general practitioner. He was prescribed azelnidipine 16 mg and rosuvastatin 5 mg. His father had had a myocardial infarction (not relevant to the history). Transthoracic echocardiography showed a large mass (approximately 35 × 37 mm) in the left atrium, and the patient was referred to our hospital for surgical resection of the large mass. At that time, he was alert and oriented, with a temperature of 37.3°C, a heart rate of 122 beats/min, blood pressure of 116/74 mm Hg, and an oxygen saturation level of 97% on room air. Diastolic heart murmur at the third left sternal border (Levine grade III/VI) and pedal pitting edema of grade 2 were found in both feet.

Chest radiography showed cardiomegaly and pulmonary congestion. Electrocardiography showed sinus tachycardia with T-wave inversion in leads V<sub>1</sub> to V<sub>3</sub>. Laboratory data showed elevated C-reactive

protein of 3.1 mg/L (normal range, <0.3 mg/L), elevated D-dimer of  $1.2 \times 10^3$  μg/L (normal range, <1.0 μg/L), and B-type natriuretic peptide of 672 pg/mL (normal range, <18.4 pg/mL).

Transthoracic echocardiography was performed at our institution. A parasternal long-axis view showed a large left atrial mass (32 × 36 × 42 mm; [Figure 1A](#), [Video 1](#)) prolapsing through the mitral valve ([Figures 1B-1 and 1B-2](#), [Video 2](#)). A short-axis view showed that the large mass was attached to the anterior and inferior portions of the left atrial wall. No protrusion to the right atrium was noted. Adhesion to the mitral valve leaflets was also suspected. There was no apparent blood flow in the mass. Continuous-wave Doppler across the mitral valve in the apical four-chamber view demonstrated a mean gradient of 26 mm Hg ([Figure 1C](#)), consistent with severe mitral stenosis. The right ventricle was dilated (basal and mid right ventricular diameters were 29 and 34 mm, respectively), and the fractional area change of the right ventricle was reduced (approximately 22%). In addition, estimated right ventricular systolic pressure was 72.3 mm Hg, suggesting severe pulmonary hypertension. Emergency surgery was consequently performed for a large mass with pulmonary hypertension and low-output status.

Transesophageal echocardiography after the introduction of general anesthesia provided additional information about the mass and indicated that the surface was rough and lobulated. The interior of the mass showed heterogeneous echo texture with few hypochoic areas ([Figure 2A](#), [Video 3](#)). The large tumor was attached to both the interatrial septum and the anterior and inferior walls of the left atrium ([Figure 2B](#), [Video 4](#)).

Intraoperative findings showed that the mass was adhered to part of the posterior mitral annulus and posterior leaflet ([Figure 3A](#)), and after adhesion dissection at the same site, the stalklike adhesion in the atrial septum was resected and excised. It was milky white, lobed, and solid ([Figure 3B](#)). The appearance of the mass was suspicious for malignancy, and intraoperative rapid pathologic examination showed malignant findings as well. The atrial septum was subsequently closed by an autologous pericardial patch. In addition, full-body positron emission tomography was performed after surgery, and it showed no lesions suggestive of malignant tumors in other organs.

The pathologic findings showed a malignant tumor in which there was proliferation of spindle cells with a large nuclear/cytoplasm ratio and necrosis on the background of a fibrotic or myxomatous stroma ([Figure 4A](#)). Strong eosinophilic cytoplasm and ubiquitously located nuclear cells with atypical cells suspected of differentiating into striated muscle were observed ([Figure 4B](#)). Immunostaining was positive for desmin and myogenin and focally positive for α-smooth muscle actin ([Figure 4C–E](#)). These findings suggested rhabdomyosarcoma, which was classified as pleomorphic, but it was not typical. Additional immunostaining showed amplification of murine double minute 2. This tumor was therefore classified as intimal sarcoma with ectopic differentiation to the striated muscle.

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Keywords: Intimal sarcoma, Primary malignant cardiac tumor, Echocardiography, Pathology, Surgery

Conflicts of interest: The authors report no conflicts of interest relative to this document.

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2468-6441

<https://doi.org/10.1016/j.case.2020.12.009>

## VIDEO HIGHLIGHTS

**Video 1:** Transthoracic echocardiography, parasternal long-axis view, demonstrating the obstruction of the mitral orifice by the large left atrial mass. **Figure 1A** corresponds to this panel.

**Video 2:** Transthoracic echocardiography, apical four-chamber view, demonstrating a mobile large left atrial mass protruding into the left ventricle. **Figure 1B** corresponds to this panel.

**Video 3:** Transesophageal echocardiography, midesophageal four-chamber view, demonstrating the rough surface and lobed large left atrial mass. **Figure 2A** corresponds to this panel.

**Video 4:** Transesophageal echocardiography, midesophageal two-chamber view, demonstrating the large left atrial mass; the interior of the mass showed heterogenous echo texture with few hypochoic areas. **Figure 2B** corresponds to this panel.

View the video content online at [www.cvcasejournal.com](http://www.cvcasejournal.com).

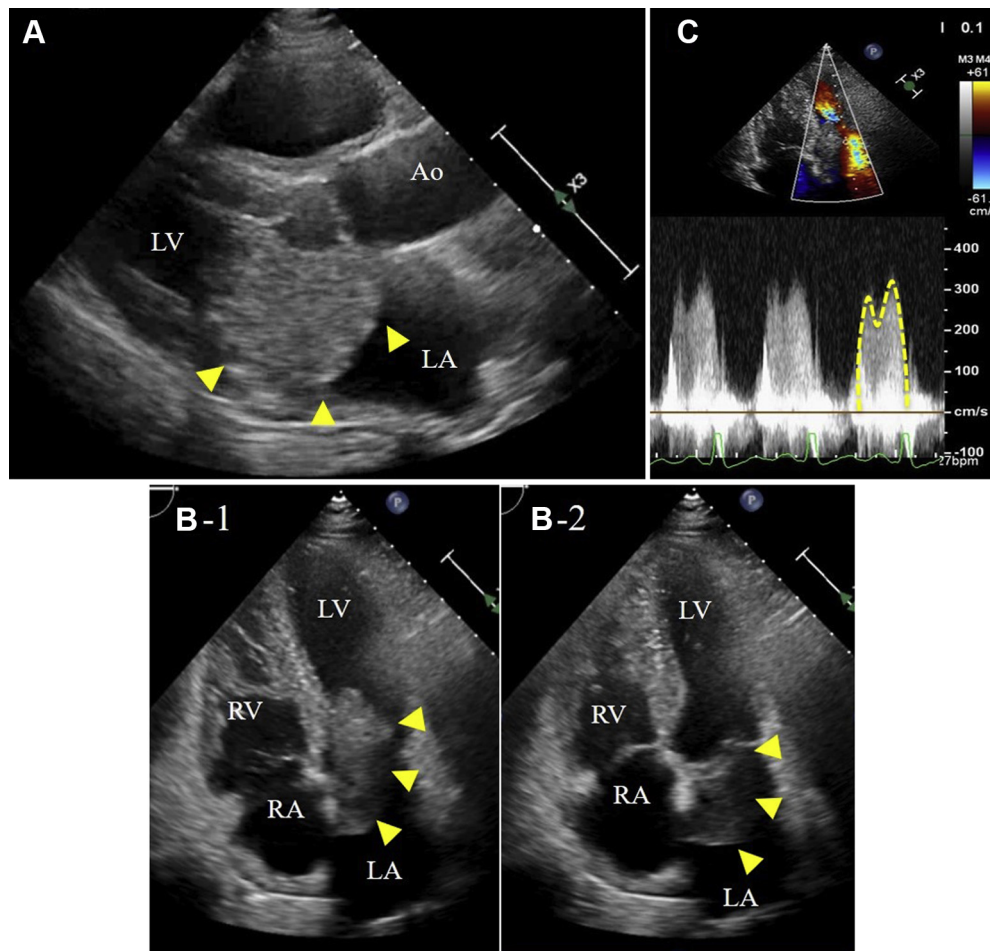
## DISCUSSION

## Primary Malignant Cardiac Tumor

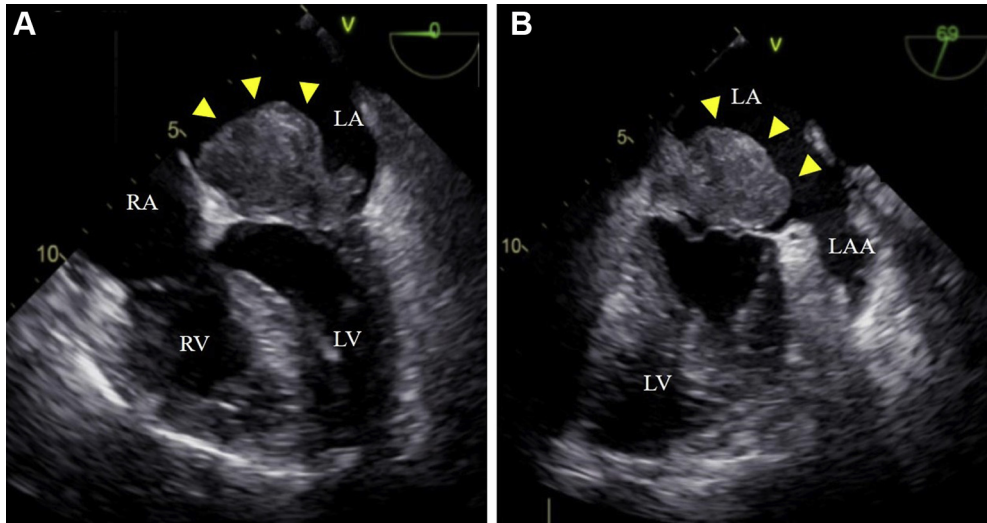
Primary malignant cardiac tumors are very rare, occurring in only 0.001% to 0.03% of patients in an autopsy series.<sup>3</sup> Kamiya *et al*<sup>4</sup> reported that in 26 of 34 patients (76%) who underwent surgical treatment, primary cardiac tumors were present in the left atria. Of the 26 left atrial tumors, 22 (84%) were myxomas, two (8%) were benign nonmyxoma tumors, and two (8%) were sarcomas.<sup>4</sup>

Occurrence Site of a Primary Malignant Cardiac Tumor:  
Pathologic Characteristics of Left Atrial Tumors

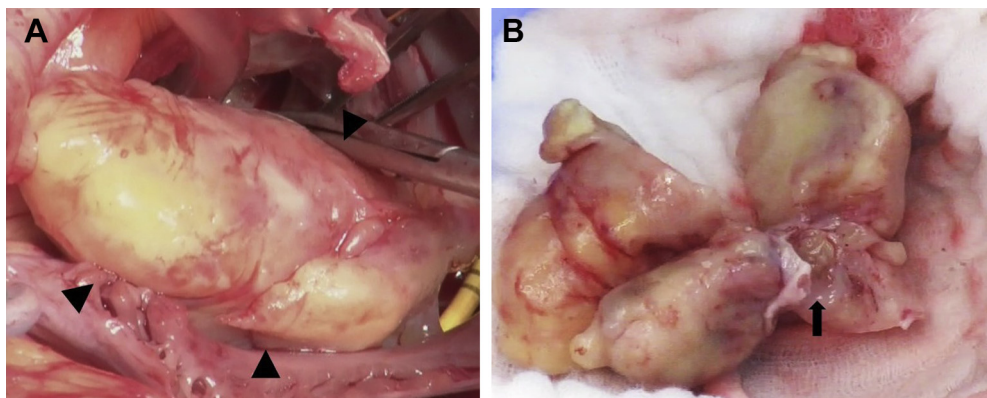
The distribution of sites and pathologic diagnoses of malignant cardiac tumors according to previous reports are shown in **Table 1**.<sup>1,5,6</sup> A tumor present only on the right side of the heart has an almost 50% chance of being a malignant primary cardiac tumor.<sup>6</sup> However, in the left atrium, undifferentiated sarcomas (24%), accompanied leiomyosarcomas (8%–9%), and osteosarcomas (3%–9%)<sup>5</sup>



**Figure 1** Images from initial transthoracic echocardiography. **(A)** Parasternal long-axis view at mid-diastole demonstrating the obstruction of the mitral orifice by the large left atrial mass (*yellow arrowheads*). **Video 1** corresponds to this panel. **(B)** Apical four-chamber view at mid-diastole (**B-1**) and systole (**B-2**) demonstrating a mobile large left atrial mass protruding into the left ventricle (LV) (*yellow arrowheads*). **Video 2** corresponds to this panel. **(C)** Continuous-wave Doppler spectrum of the mitral valve showed elevated flow velocity, and the mean pressure gradient was 26 mm Hg (*yellow dotted line*), consistent with severe mitral stenosis. Ao, Aorta; LA, left atrium; RA, right atrium; RV, right ventricle.



**Figure 2** Images from intraoperative transesophageal echocardiography. **(A)** Midesophageal four-chamber view at mid-systole demonstrating the rough surface and lobed large left atrial mass (yellow arrowheads). *Video 3* corresponds to this panel. **(B)** Midesophageal two-chamber view at mid-systole demonstrating the large left atrial mass; the interior of the mass showed heterogenous echo texture with few hypoechoic areas (yellow arrowheads). *Video 4* corresponds to this panel. LA, Left atrium; LAA, left atrial appendage; LV, left ventricle; RA, right atrium; RV, right ventricle.



**Figure 3** Images from intraoperative pictures of the mass. **(A)** Intraoperative picture seen from the left atrium showing the mass (black arrowheads) with adhesion to the posterior mitral annulus (black arrow). **(B)** Excised specimen showing the milky white and lobed large mass. Black arrow denotes the stalklike adhesion to the atrial septum.

are the most likely. Intimal sarcomas are typically tumors of the great vessels (pulmonary artery, aorta, pulmonary veins, and vena cava). A possible link has been suggested between left atrial sarcomas and intimal sarcomas because of their intracavitary growth. However, Neuville *et al*<sup>7</sup> reported that intimal sarcoma was the most common type upon reclassifying cardiac tumors that exhibit murine double minute 2 amplification and are often located in the left heart.

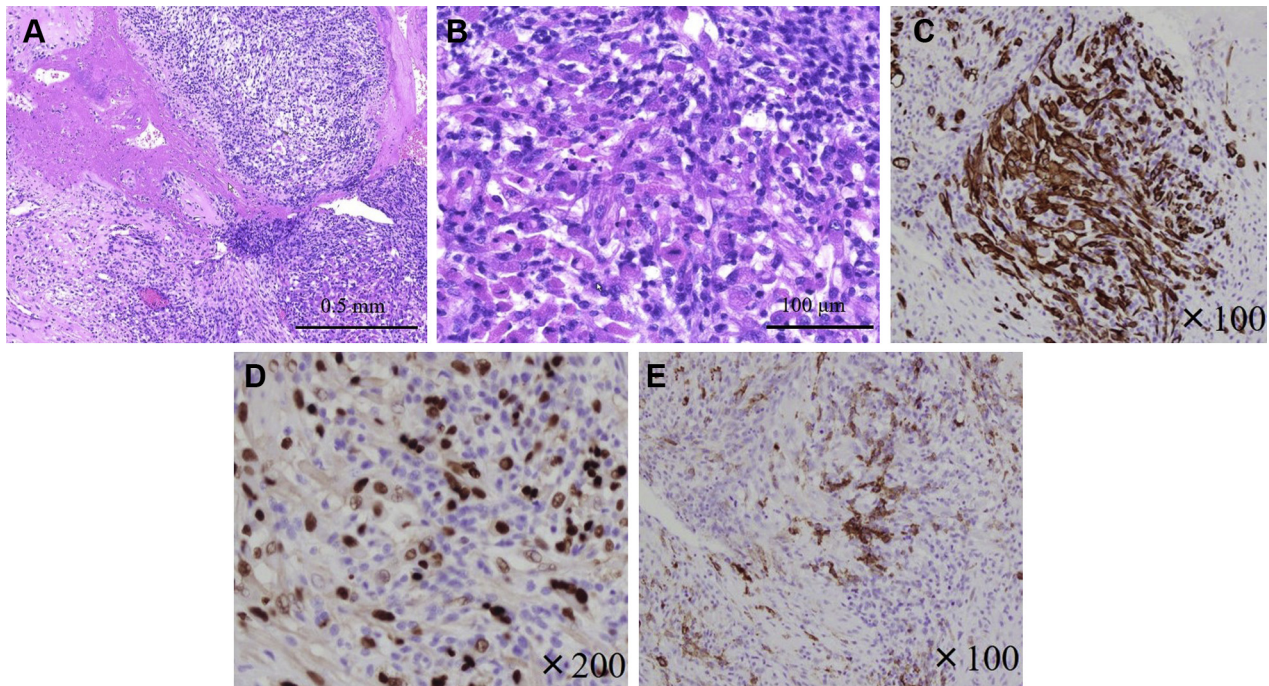
#### Echocardiographic Findings of Left Atrial Tumors: Difference between Left Atrial Myxoma and the Present Case

Cardiac masses are usually differentiated by clinical context, anatomic location, and appearance on echocardiography. Myxomas are usually heterogeneous, pedunculated, mobile tumors with a broad-based endocardial attachment. Most atrial myxomas arise from the interatrial septum in the region of the fossa ovalis.<sup>3</sup> They may be homoge-

neous or may have central areas of hyperlucency representing hemorrhage and necrosis.<sup>3</sup> However, this case showed a rough surface and a lobed mass. The interior of the mass showed a heterogeneous echo texture. The large tumor was attached to both the interatrial septum as well as the anterior and inferior walls of the left atrium. However, myxoma with a broken capsule may also present a rough surface with lobed features; therefore, differentiation by echocardiography may be difficult.

#### CONCLUSION

Cardiac intimal sarcoma is very rare, and early diagnosis using echocardiography is important but remains challenging. This case adds to the limited literature with cardiac intimal sarcoma as one of the differentiations of left atrial tumors.



**Figure 4** (A) Hematoxylin and eosin staining of weak expansion showing proliferated spindle cells with a large nuclear/cytoplasm ratio. Necrosis was also seen in the background of a fibrous or myxomatous stroma. (B) Hematoxylin and eosin staining of strong expansion showing that the cells contained strong eosinophilic cytoplasm and a ubiquitously located nucleus. (C) Immunostaining image showing positivity for desmin. (D) Immunostaining image showing positivity for myogenin. (E) Immunostaining image showing focal positivity for  $\alpha$ -smooth muscle actin.

**Table 1** The distribution of primary malignant cardiac tumors: site and pathologic diagnosis

	Site					
	Left atrium	Left ventricle	Right atrium	Right ventricle	Pericardium	Blood vessels
Pathologic diagnosis	Undifferentiated sarcoma	Lymphoma	Undifferentiated sarcoma	Angiosarcoma	Liposarcoma	Undifferentiated sarcoma
	Leiomyosarcoma		Angiosarcoma	Liposarcoma	Lymphoma	Intimal sarcoma
	Osteosarcoma		Leiomyosarcoma	Lymphoma		Leiomyosarcoma
	Rhabdomyosarcoma		Synovial sarcoma			
	Fibrosarcoma		Liposarcoma			
	Fibrous histiocytoma		Lymphoma			
	Lymphoma					

## ACKNOWLEDGMENTS

We thank H. Ikeda for useful comments on pathology and A. Nishiyama and H. Tada for advice and discussions.

## SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2020.12.009>.

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