Clinical Case Report

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# A case of a resected benign myxoma-like hemorrhagic cyst, which later recurred as undifferentiated pleomorphic sarcoma in the left atrium

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**Abstract**

**Rationale:** An intracardiac cystic mass is a rare type of mass found in the left atrium. The differential diagnosis of an intracardiac cystic mass includes hydatid cysts, bronchogenic cysts, intracardiac varices, and hemorrhages in some tumor types, including myxoma.

**Patient concerns:** We present the case of a 68-year-old woman who presented with episodic dyspnea.

**Diagnoses-Interventions-Outcomes:** Transthoracic echocardiography (TTE) revealed the presence of a left atrial mass mimicking myxoma. However, in postoperative findings, it was determined that the mass was actually a hemorrhagic cyst. Eighteen months later, the patient presented with recurrent exertional dyspnea and TTE revealed the recurrence of a left atrial mass. Computed tomography showed that the mass extended into the right atrium, inferior vena cava, and coronary sinus. After re-operation, the final histological diagnosis was determined to be an undifferentiated pleomorphic sarcoma in the left atrium.

**Lessons:** An intracardiac hemorrhagic cyst was suspected during the operation of a benign-looking LA mass. As such, we recommend that other rare etiologies be considered and more biopsies be performed when possible.

**Abbreviations:** CT = computed tomography, LA = left atrium, MRI = magnetic resonance imaging, TTE = transthoracic echocardiography.

**Keywords:** cardiac sarcoma, hemorrhagic cyst, left atrial mass

## 1. Introduction

An intracardiac cystic mass is a rare type of mass found in the left atrium. The differential diagnosis of an intracardiac cystic mass includes hydatid cysts, bronchogenic cysts, intracardiac varices,[1] and hemorrhages in tumor types (e.g., myxoma).[2] We present a case of left atrial (LA) undifferentiated pleomorphic sarcoma in which the initial presentation was an intracardiac hemorrhagic cyst mimicking clinical LA myxoma.

## 2. Case report

A 68-year-old woman complained of episodic dyspnea and chest discomfort over 2 months while walking. These symptoms typically lasted for several minutes and resolved spontaneously. Transthoracic echocardiography (TTE) showed the presence of severe functional mitral valve stenosis and moderate mitral regurgitation, a large round echogenic mass attached to the LA side of the posterior mitral valve leaflet (Fig. 1A), and severe pulmonary hypertension. Transesophageal echocardiography demonstrated that a well-capsulated echogenic mass ($36 \times 15$ mm) occupied the superoposterior half of the left atrium and extended to the posterior mitral valve leaflet (Fig. 1B). The patient's clinical diagnosis was LA myxoma; as such, we decided to perform an en-block resection of the LA mass. After the first operation, we observed that the excised specimen was a bag-like cystic mass $4.2 \times 2.9 \times 1.1$ cm in size, which contained a large amount of dark brown to red-colored coagulated blood (Fig. 1C). Pathologic findings showed that the atrial wall had mixed thickening with fibrosis and a blood cyst in the endocardium with mild, chronic, and active inflammatory changes (Fig. 1D). To determine the etiology of the blood cyst, we performed additional random biopsies of the left atrium. Through pathological assessment, we found that these

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*EK and S-WC contributed equally this study.*

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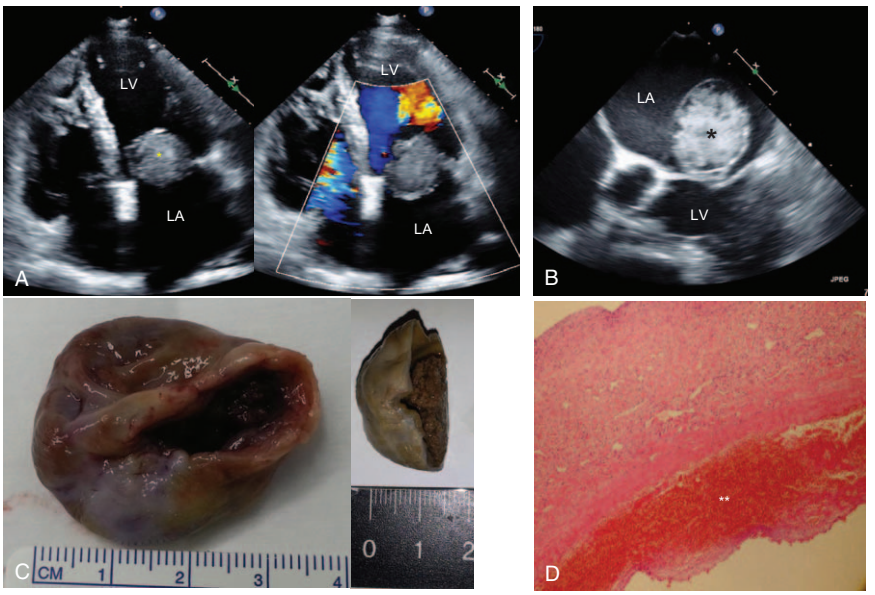


**Figure 1.** (A) Transthoracic echocardiography showing a left atrial mass (*) and functional mitral stenosis. (B) Transesophageal echocardiography. (C) After resection, the excised specimen consisted of a bag-like cystic mass (4.2 × 2.9 × 1.1 cm in dimension), which contained a large amount of dark brown to red-colored coagulated blood (**). (D) Hematoxylin and eosin (H&E), ×400. LA = left atrium, LV = left ventricle.

samples were normal endocardium. Our final diagnosis was an LA intracardiac hemorrhagic cyst mimicking myxoma, and the patient was discharged without any complications.

One and a half years later, the patient complained of recurrent exertional dyspnea and presented with a respiratory rate of 18/min, a heart rate of 74 bpm, and blood pressure of 110/80 mm Hg. A cardiovascular exam showed a low-pitched, grade 3/6 diastolic rumbling murmur at the apex. Lung sounds were clear with no wheezing, and the patient's abdomen was soft with normal bowel sounds.

Laboratory findings were within normal limits, with the exception of the concentration of N-terminal probrain natriuretic peptide (2283 pg/mL). An electrocardiogram showed normal sinus rhythm with T-wave inversion in the V1–V3 leads. A chest x-ray showed mild pulmonary edema and small amounts of bilateral pleural effusion. TTE revealed a large round echogenic mass attached at the left atrium (Fig. 2) and severe functional mitral valve stenosis. Cardiac computed tomography (CT) and heart magnetic resonance imaging (MRI) showed a mass of 82 × 62 mm with irregular margins, heterogeneous enhancement in the

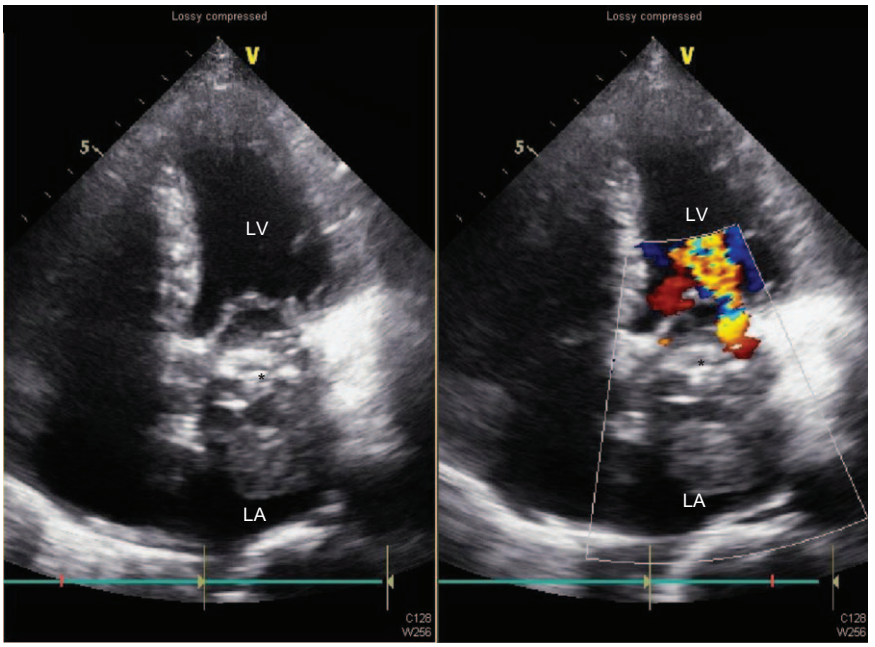


**Figure 2.** Eighteen months after left atrial (LA) mass (*) excision, a recurred LA mass, and functional mitral stenosis were observed by echocardiography. LA = left atrium, LV = left ventricle.

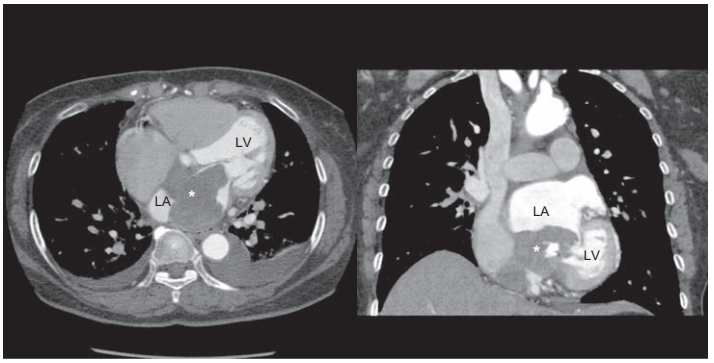


**Figure 3.** Computerized tomography demonstrated that the mass (*) in the left atrium had irregular margins and extended to both the right atrium and coronary sinus. LA = left atrium, LV = left ventricle.

left atrium, and extensions into the right atrium, inferior vena cava, and coronary sinus (displacement rather than invasion) (Figs. 3 and 4). Positron emission tomography/CT revealed no visible metastatic lesions (Fig. 5).

The patient was referred for surgery and cardiac exploration was planned. Before her operation, coronary angiography was performed and no coronary artery stenosis or vascular malformations were discovered. The patient underwent resection of the LA tumor, as well as an LA wall biopsy. The excised specimens were irregularly shaped, consisted of pinkish tan-colored soft tissue and partly yellowish myxoid, and were partially fibrous without hemorrhage or necrosis. Pathological assessment showed that the specimens had hyperchromatic spindle cells in the myxoid background with prominent arborizing vessels and both undifferentiated pleomorphic and giant cells (Fig. 6). Our final diagnosis was LA undifferentiated pleomorphic sarcoma.

## 3. Discussion

Primary cardiac tumors are rare. Three-quarters of these tumors are benign, and nearly half of the benign tumors are myxoma. Overall, 25% of cardiac tumors are malignant, and, of these, 95% are sarcomas (the most common of these are undifferentiated). The remaining types of tumors are comprised of mostly angiosarcomas, leiomyosarcomas, and rhabdomyosarcomas. Notably, the most common malignancy affecting the heart is secondary from other organs.[3] Undifferentiated pleomorphic sarcoma, previously known as pleomorphic malignant fibrous histiocytoma, is lobulated and can be sessile or pedunculated, often reaching 10 cm in diameter. These sarcomas are most commonly found in the left atrium, with patients experiencing pulmonary congestion and hemopericardium. This tumor type is sometimes misdiagnosed as myxoma.[4]

As reported previously, there are several findings of LA sarcomas that distinguish them from benign myxomas:[5] (1) a mass of nonseptal origin, (2) extension into the pulmonary vein, (3) multiple masses, (4) a broad attachment on the LA wall, and (5) a semisolid consistency. There are rare case reports of primary cardiac sarcomas that present preoperatively as benign myxomas,[6,7] although echocardiography and imaging modalities

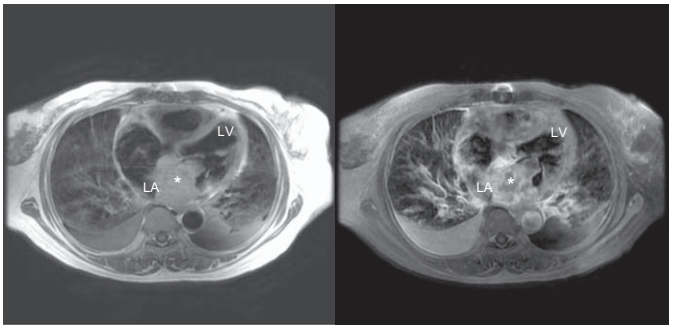


**Figure 4.** The mass (*) in the left atrium showed high signal intensity on T1-weighted magnetic resonance images. The mass extended to the right atrium, coronary sinus, and inferior vena cava. LA = left atrium, LV = left ventricle.

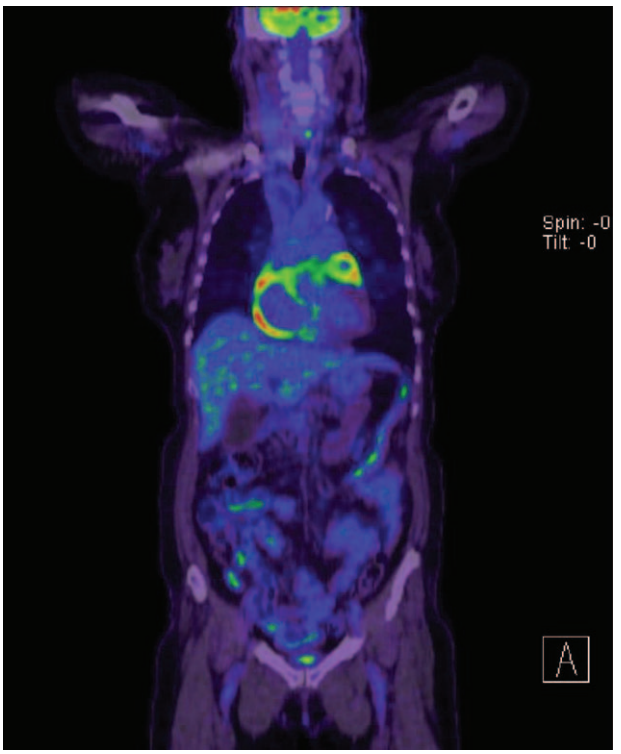


**Figure 5.** Positron emission tomography/computerized tomography showed intensive radioactive uptake around both atria and no distant metastasis.

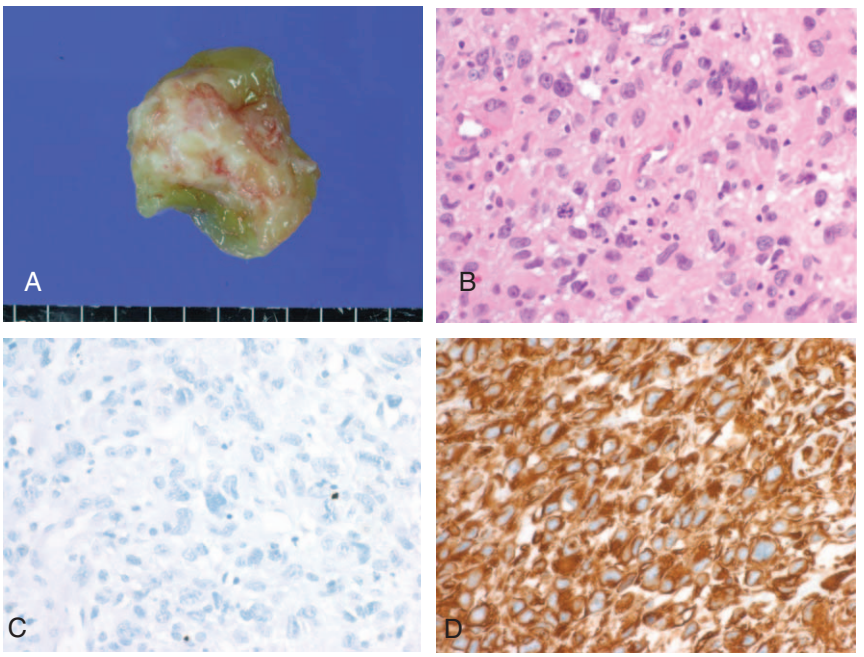


**Figure 6.** (A and B) Gross and microscopic findings. Histologically, the undifferentiated pleomorphic sarcoma expressed highly pleomorphic epithelioid cells with numerous mitotic and interspersed giant cells arranged haphazardly in a sheet (H&E, × 400). (C) S-100 staining: negative. (d) Vimentin staining: positive. H&E = hematoxylin and eosin.

(e.g., MRI or CT) can aid in their diagnosis. Regardless, the precise differential diagnosis of an LA mass may not always be easy, especially in rare cases with atypical morphologic characteristics.

Although most cases of myxoma appear as round and solid masses without cystic architecture, several cases of cystic myxoma have been reported. To our knowledge, we are the first to report an intracardiac hemorrhagic cyst without a tumor and other predisposing factors. Therefore, we studied both resected tissue and additional random LA biopsies and found only normal endocardium. After the first operation, we determined that the mass was an intracardiac hemorrhagic cyst mimicking myxoma. After reoperation, we determined that the mass was indeed an undifferentiated pleomorphic sarcoma or hidden sarcoma, which was not detected initially.

In conclusion, we believe that our case represents the first case of LA undifferentiated pleomorphic sarcoma in which the initial presentation was an intracardiac hemorrhagic cyst mimicking myxoma. According to our case, an intracardiac hemorrhagic cyst was suspected during the operation of a benign-looking LA mass. As such, we recommend that other rare etiologies be considered and more biopsies be performed when possible.

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