

Case Report

Cardiac myxoma and its growth over 5 years in a patient with a history of multiple cerebrovascular events: a case report

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ABSTRACT

Cardiac tumors frequently present significant clinical, diagnostic, and therapeutic challenges. In many cases, a cardiac mass is detected incidentally and can represent benign or malignant processes, primary or secondary. Over 90% of primary cardiac tumors are benign (in adults, myxomas are most common, while in children, rhabdomyomas are more frequent). These tumors can be further classified as simple or complex based on the necessary treatment and may affect different chambers of the heart and the pericardium. Secondary or metastatic cardiac tumors are 30 times more common than primary neoplasms, with an incidence in autopsies ranging from 1.4% to 14%. Myxomas constitute approximately 50% of all benign cardiac tumors in adults. Most myxomas (over 80%) are located in the left atrium, with decreasing frequency in the right atrium, right ventricle, and left ventricle. We present the case of a 76-year-old woman who was found to have a left atrial mass identified on echocardiogram in 2019 as part of a cerebrovascular disease evaluation protocol. However, the patient did not attend follow-up. In December 2024, she was hospitalized for a lower respiratory tract infection, and during her admission, a new echocardiographic study showed tumor growth over 5 years.

Keywords: Cardiac tumor, Myxoma, Cardiac tumors

INTRODUCTION

Cardiac myxomas are the most common primary heart neoplasms. They can affect any chamber of the heart however, the left atrium is the most frequently involved, in 75% of cases, followed by the right atrium (15-20%) and the left and right ventricles (3-4%).¹ They tend to develop in the interatrial septum and the fossa ovalis, though they may also extend from the anterior or posterior atrial wall. These features are typically present in solitary and sporadic myxomas.² Myxomas arising from sites other than the left atrium or those that are multicentric should

always raise suspicion for carney complex, a syndrome resulting from germline mutations in PRKAR1A (a gene that encodes the regulatory subunit alpha-1 of protein kinase A located at loci 17q22-24 and 2p16).³

Immunohistochemical detection of PRKAR1A is useful for diagnosis and can guide detection in patients and their families.⁴ Macroscopically, myxomas appear yellow, white, or brownish, and are often friable.⁵ Microscopically, they are composed of spindle-shaped and stellate cells (myxoma cells) with a myxoid stroma, which may also contain endothelial cells, smooth muscle cells,

and other elements within a mucopolysaccharide substance. Although they may be discovered incidentally during imaging studies, nearly 70% of patients with myxomas present significant symptoms such as stroke, peripheral embolization, and sudden death. Constitutional symptoms are also common, affecting up to a third of patients, and can include fever, weight loss, fatigue, myalgias, arthralgias, and even Raynaud's phenomenon.

Other symptoms may include positional dyspnea, chest pain, congestive heart failure, edema in the limbs and neck, palpitations, arrhythmias, pericardial effusion, or tamponade.⁶ These clinical manifestations are summarized in Table 1.

Knowledge of the distribution of cardiac masses according to anatomical location (Table 2) by imaging methods enables accurate diagnosis and treatment planning without the need for open or percutaneous surgical biopsy.⁷ Imaging tests should assess the possibility of cardiac surgery.⁸

Transthoracic echocardiography is usually the first diagnostic choice, providing high-quality dynamic images that can be performed at the patient's bedside. It is widely used for morphological and functional heart assessment.⁹

It allows evaluation of the tumor size, location, mobility, and pericardial involvement. Despite its good spatial resolution and utility in understanding the hemodynamic significance of the mass, its limitations include poor acoustic windows (in obese patients or those with COPD), limited tissue characterization, and occasional difficulty in distinguishing the extent and origin of the mass.²

In cases where transthoracic echocardiography is insufficient due to suboptimal image quality, contrast echocardiography may improve the image quality and help differentiate benign from malignant lesions. A prospective cohort study across four tertiary hospitals in China, involving 153 patients with suspected cardiac masses, demonstrated notable diagnostic performance of contrast

echocardiography, with high sensitivity and specificity for distinguishing cardiac tumors from non-neoplastic cardiac masses and comparable accuracy to pathological analysis in differentiating malignant from benign tumors. Ultrasound contrast agents are useful in the differential diagnosis of thrombi, for visualizing hypoechoic masses in the atria, and for enhancing Doppler signals in highly vascularized lesions.¹⁰

Magnetic resonance imaging (MRI) is employed for tissue characterization of cardiac tumors and has greater sensitivity and specificity in detecting cardiac masses and in differential diagnosis. Several parameters can be considered, including inversion time, first-pass perfusion, and late gadolinium enhancement post-contrast.¹¹ In cardiac MRI, myxomas appear as heterogeneous masses on various T1 and T2-weighted sequences pre- and post-gadolinium contrast due to varying amounts of myxoid, haemorrhagic, calcified, and necrotic tissue.¹²

Heterogeneous gadolinium late enhancement may be seen, along with non-homogeneous lesions, hypo intensity or is intensity on T1-weighted images, and hyperintensity on T2-weighted sequences.¹³ Regarding the multiplicity of myxomas, they have a well-established potential to metastasize. Literature reports have found myxomas in various organs, including the pancreas, kidneys, stomach, bones, and even the skin.

Myxomas may contain variable portions of calcifications (10-20% of myxomas) and post-necrotic and post-haemorrhagic cysts. Additionally, the vascularization of these tumors is highly variable.

Most are hypo vascular and show weak enhancement with contrast medium, while others are hyper vascular and may even contain a fistula between coronary arteries and cardiac chambers. Antoniac et al presented a case of a highly vascularized myxoma with a possible fistula between the branches of the left circumflex artery and the left atrium.¹³

Table 1: Clinical characteristic of cardiac myxoma.

Clinical characteristic	
Constitutional signs and symptoms	Fever, weight loss, general malaise, anorexia, fatigue and arthralgia.
Embolic signs and symptoms	Depends on the site of embolization: cerebrovascular accident, transient ischemic attack, acute ischemia of the extremities, viscera; spleen, adrenal glands, kidneys and abdominal aorta. Raynaud's phenomenon, abdominal pain, diarrhea.
Obstructive cardiac symptoms and hemodynamic consequences	Dyspnea, intracardiac obstruction, pseudo-obstruction of the mitral valve, pulmonary hypertension, syncopal episodes due to valve obstruction, palpitations, conduction abnormalities, arrhythmia, heart failure when right atrial myxomas obstruct the tricuspid valve.
Atypical presentation	Fever of unknown origin, repetitive dysphagia, hemoptysis, pleural effusion, acute myocardial infarction.

Table 2: Location of primary and secondary cardiac tumors.

Location of primary and secondary cardiac tumors	
Right atrium	Lipoma, lymphoma, metastasis, myxoma, Sarcoma.
Left atrium	Lipoma, metastasis, myxoma, sarcoma.
Left ventricle	Fibroma, lipoma, lymphoma, metastasis, rhabdomyoma, sarcoma.
Right ventricle	Fibroma, lipoma, lymphoma, metastasis, rhabdomyoma.
Inferior vena cava	Leiomyoma, kidney tumor.
Pulmonary veins	Lung tumors, sarcoma.
Pulmonary artery	Sarcoma.
Valves	Fibroelastoma, metastasis.
Pericardium	Liposarcoma, lipoma, lymphoma, mesothelioma, metastasis.

Left atrial myxomas are considered highly curable with excellent long-term results. Incomplete resection, multifocal tumors, and tumor embolism are important factors for recurrence. Metastases can be explained by the shedding of the friable, spongy component of the atrial myxoma into the systemic circulation.¹⁴ Urgent surgical intervention is indicated in symptomatic patients, and rapid surgical intervention is warranted in asymptomatic patients with polypoid myxomas to prevent embolism.¹⁵

CASE REPORT

We present the case of a 76-year-old woman with a history of three cerebrovascular events, the first in October 2018, the second in November 2018, and the last in April 2019. The patient underwent diagnostic workup for the 2019 stroke, including Doppler carotid ultrasound, which showed no stenosis or hemodynamic impact.

A 24-hour Holter study was also performed without detecting rhythm or conduction disorders. A transthoracic echocardiogram revealed a left atrium with no evidence of dilation (volume of 35 ml), and a mass attached to the

interatrial septum measuring approximately 23×22 mm (Figure 1 and 2). The patient was scheduled for follow-up by cardiology, but she did not return for further evaluation of the mass.

In November 2024, the patient was admitted to our hospital's emergency department with respiratory symptoms, Vital signs, blood pressure 100/60 mmHg, heart rate 90 bpm, respiratory rate 28, Temperature 100.9°F, developing acute kidney injury and requiring renal replacement therapy, including peritoneal dialysis catheter placement.

During her hospitalization, she completed antimicrobial treatment with ureidopenicillin, and a new echocardiographic study revealed a left atrium with no dilation (volume of 35 ml), and a mass in the left atrium attached to the septum, now measuring 40×36 mm (Figure 3 and 4), showing tumor growth since 2019. The patient experienced complications with probable peritonitis associated with peritoneal dialysis, developed hospital-acquired pneumonia, progressed to septic shock, and subsequently passed away.



Figure 1: Transthoracic echocardiogram from May 3, 2019. The patient has a poor acoustic window in the five-chamber apical view. A regular-bordered echogenic mass is observed in the left atrium. The mass measures approximately 2.3 cm×2.1 cm, with an approximate area of 4.5 cm². The lesion originates in the region of the interatrial septum near the mitral valve and extends into the left atrial cavity.

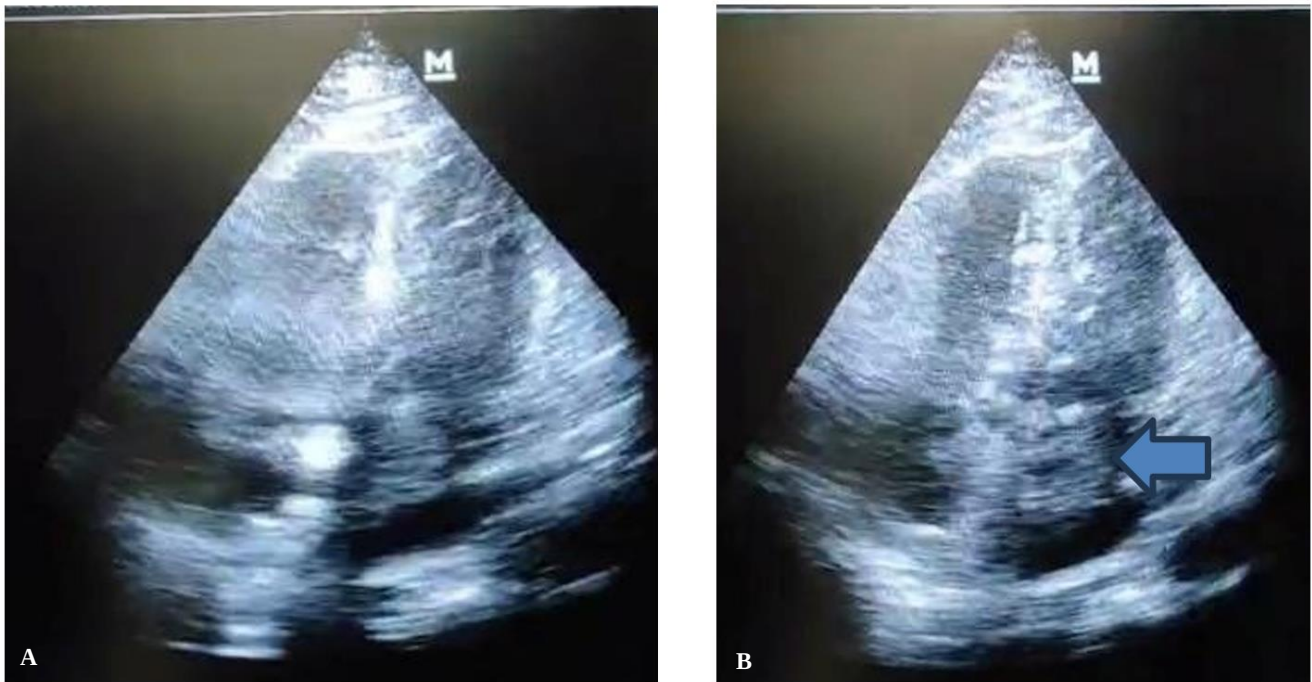


Figure 2 (A & B): The patient has a poor acoustic window in the four-chamber apical view. An echogenic mass with irregular borders is observed in the left atrium. The mass measures approximately 4.4 cm × 3.4 cm, with an approximate area of 7.8 cm². The lesion originates in the region of the interatrial septum near the mitral valve and projects into the left atrial cavity. The mass is mobile with the cardiac cycles, has a pedicle that is difficult to characterize, and displaces the mitral valve.

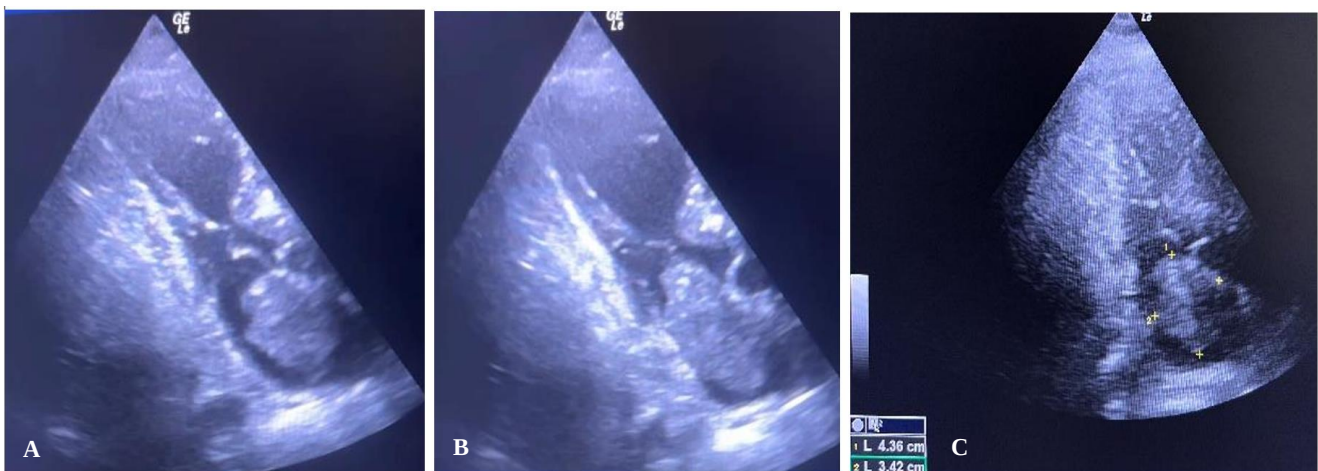


Figure 3 (A-C): December 2024, parasternal window in the long axis with vertical projection. An echogenic mass with irregular borders is observed in the left atrium. The mass measures approximately 4.36 cm x 3.42 cm, with an approximate area of 7.78 cm². The lesion originates in the region of the interatrial septum near the mitral valve and projects into the left atrial cavity, occupying more than 80% of the cavity. The mass is mobile with the cardiac cycles, has a pedicle that is difficult to characterize, and displaces the mitral valve during both the systolic and diastolic phases of the atrium.

DISCUSSION

Cardiac myxomas are difficult to detect due to the lack of specific signs and symptoms, even though benign tumors can lead to severe consequences.¹ The clinical context provides crucial diagnostic clues to establish the etiology of a mass or lesion. The size of the tumor can range from

1 cm to over 10 cm, with a smooth surface in most cases. Typically, they form a pedunculated mass with a short, broad base but can also present as sessile. A fibrous or papillary form of the myxoma has been reported, with a surface consisting of multiple fine, gel-like, fragile extensions that tend to fragment spontaneously and are associated with embolic phenomena.¹⁶

Our patient presented multiple cerebrovascular events that may have been causally related to the cardiac mass she had. She denied constitutional symptoms or other related symptoms. The echocardiographic report described the mass as measuring 40×36 mm with smooth borders and a narrow pedicle attached to the interatrial septum, reaching the mitral ring during diastolic motion.

Transthoracic echocardiography is the optimal imaging diagnostic modality for obtaining images of masses smaller than 1 cm, making it a suitable tool for evaluating these tumors. The average growth rate of cardiac myxomas is reported to be 0.49 cm/month, but the actual growth rate remains controversial.¹ Echocardiographic studies are operator-dependent, but a substantial difference in tumor size can be seen when comparing the transthoracic echocardiograms performed on our patient from the initial diagnosis in May 2019 to December 2025 (45 months). The tumor grew from 5.57 cm³ in 2019 to 24.42 cm³ in 2024, a growth rate of 0.41 cm³/month.

We chose to present this case because it describes a patient with three cerebrovascular events in the context of a left atrial cardiac mass, which was characterized as a myxoma based on clinical and echocardiographic features, and which continued to grow over the years.

CONCLUSION

Left atrial myxomas, although rare, are particularly relevant in patients over 80 years old, where overlapping symptoms with cardiovascular and neurodegenerative conditions of aging or comorbidities may complicate the diagnostic suspicion. The clinical presentation can be atypical: progressive fatigue, exertional dyspnea, or sudden cognitive decline often attributed to heart failure or dementia-delaying the suspicion of a myxoma. However, it is vital to recognize the persistent embolic risk and the increased likelihood of ischemic brain events. This case highlights that in elderly patients with a history of cerebrovascular events without a defined etiology or sudden functional deterioration, myxoma should be included in the differential diagnosis, prioritizing the use of diagnostic methods such as transoesophageal echocardiography.

Surgery remains the cornerstone of therapy, with acceptable survival rates when performed in experienced centres, considering factors like frailty, physiological reserve, and patient expectations, which influence the surgical decision. This case emphasizes the importance of suspicion for this entity and the need for appropriate multidisciplinary management and timely diagnosis to prevent short-, medium-, and long-term complications.

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