

Left Atrial Myxoma with Ischemic Stroke Complications: A Case Report

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Left Atrial Myxoma with Ischemic Stroke Complications: A Case Report

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Abstract. Cardiac myxoma is the most frequent primary tumor of the heart, considered a benign, slowly proliferating neoplasm. The incidence of cardiac myxoma is low, with approximately 0.5–1 cases per 1,000,000 individuals per year. Cardiac myxoma can be present at any age but most often in 30–60 years old, with female predominance (1.5:1). Serious complications include neurological symptoms, heart failure, arrhythmias, and pericardial effusion. Ischemic stroke complications associated with atrial myxoma due to embolus. Aims: to describe the features, complications and management of left atrial myxoma. Case summary: A 31-year-old woman with the chief complaint of sudden diplopia and syncope since 4 days before admission to the hospital. Patient also had history of recurrent vertigo in the last month. Her vital sign was normal. Neurological examination showed eyeball muscles paralysis (OD: nerve VI, IV and III inferior oblique paralysis; OS: nerve III medial and superior paralysis), flat right nasolabial plica (nerve VII right perifer paralysis) and tongue deviation to the right (nerve XII right central paralysis). A Head MRI showed subacute multiple infarcts in bilateral thalamus and bilateral centrum semiovale with infarcts in left cerebellum. Echocardiography showed a pedunculated mass in left atrium attached to the interatrial septum, which was suggestive of left atrial myxoma. Patient diagnosed a left atrial myxoma with ischemic stroke. Results: Excised left atrial myxoma was performed with cardiac surgery and cardiopulmonary bypass procedures. After treatment procedures, the patient have clinical improvement without any complications. Histopathological examination revealed cardiac myxoma. Follow-up echocardiography showed no residual tumors in left atrial. Conclusions: Echocardiography should be done in all cases of stroke as a screening tool to rule out any intracardiac mass, such as cardiac myxoma. Early diagnosis and urgent surgical excision of the cardiac myxoma remains the definitive treatment that will be given excellent results.

Keywords: myxoma · echocardiography · ischemic stroke · cardiac surgery

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1 Introduction

Cardiac myxoma is the most common primary tumor of the heart, as a benign slowly proliferating neoplasm of endocardial origin [1–3]. The incidence of cardiac myxoma is low, about 0.5–1 cases per 1,000,000 people per year [2]. Myxoma can be present at any age [1] but most often between the 30–60 years of age, [4] with a slight female predominance (1.5:1) [1]. Clinical presentation of cardiac myxomas varies with non-specific cardiac symptoms, depends on the size, location, propensity for embolization, invasiveness, and relation with other cardiac structures [1]. Clinical symptoms due to mitral valve obstruction may cause syncope, dyspnea, and pulmonary edema followed by embolic manifestations [1, 2]. Moreover cardiac myxoma may be symptomatic present or found incidentally during evaluation for a seemingly unrelated problem or physical findings [1]. An echocardiographic examination should be done to establish intracardiac mass, such as cardiac myxoma [1]. Serious complications include neurological symptoms (stroke), heart failure, arrhythmias, pericardial effusion [4], and sudden death [5]. Ischemic stroke complications associated with atrial myxoma due to embolus [6]. Embolic events due to detached tumor tissue, disseminated thrombotic material overlying the tumor, or a compound of both, have been shown to occur in up to 30–50% of patients [2]. The location of cardiac myxoma is often in the left atrium (75–90%) so that more than 50% of embolic events affect the central nervous system and the retinal arteries [2].

The definitive treatment of cardiac myxoma is immediate cardiac surgery for myxoma mass resected with cardiopulmonary bypass (CPB) and cardioplegic procedures [1, 3, 4, 7] (Table 1).

Table 1. TIMELINE

Time	Event
Day 1	The patient presented with complaints of sudden diplopia and syncope since 4 days ago. The patient also had recurrent vertigo in the last month. The ECG and chest X-ray were normal. A head CT scan showed right and left thalamus infarcts,
Day 6	Echocardiography showed a pedunculated mass attached to the left inter-atrial septum and was suspected as a left atrial myxoma. A head MRI showed subacute multiple infarcts in the bilateral thalamus and bilateral centrum semiovale with infarcts with the left cerebellum.
Hari 8	The patient underwent open-heart surgery via median sternotomy and cardiopulmonary bypass procedures. The left atrial myxoma was resected and the interatrial septum defect was repaired. The patient was monitored closely after operation in the cardiovascular care unit.
Hari 12	The patient's condition improved clinically without any complications. Follow-up echocardiography showed good left ventricular function and no residual tumor mass in the atrial left.
Hari 16	The patient was a follow-up in the out-patient.

2 Case

A 31-year-old woman presented to the emergency department with the chief complaint of sudden double vision (diplopia) and syncope 4 days before admission to the hospital. The patient also often complains of recurring dizziness (vertigo) associated with nausea and vomiting that resolved within hours in the past month. She has no history of shortness of breath, chest pain, and hypertension. She has no family history of the tumor. On admission, she was hemodynamically stable, with normal blood pressure (110/70 mmHg), pulse 86 beats/minute, respiratory rate 20 times/minute, temperature 36.5 °C. Auscultation of her chest revealed a regular rate with normal heart sound and no murmur. Neurological examination revealed the eyeball muscles paralysis (OD: nerve VI, IV, and III inferior oblique paralysis; OS: nerve III medial and superior paralysis), flat right nasolabial plica (nerve VII right perifer paralysis), and tongue deviation to the right (nerve XII right central paralysis).

Laboratory finding was dyslipidemia (LDL 164 mg/dL, HDL 45 mg/dL, total cholesterol 215 mg/dL, Triglycerides 151 mg/dL) and the other were within normal limits. An electrocardiogram showed normal sinus rhythm. Chest X-ray was normal limits. A head computed tomography (CT) scan showed infarcts of the right and left thalamus, especially the right. Magnetic resonance imaging (MRI) of her brain demonstrated subacute multiple infarcts in the bilateral thalamus and bilateral centrum semiovale with infarcts with blood components in the left cerebellum (Fig. 1). Transthoracic echocardiography

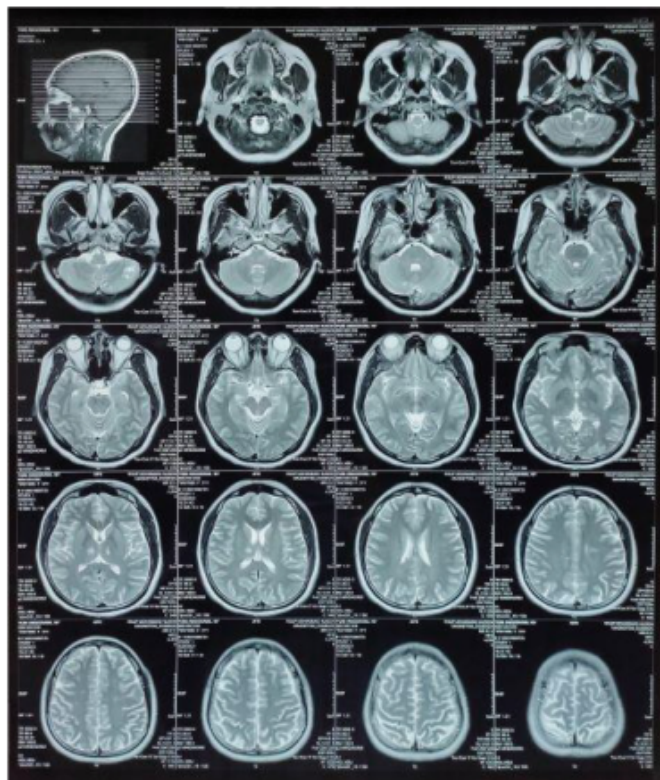


Fig. 1. A head MRI showed subacute multiple infarcts in the bilateral thalamus and bilateral centrum semiovale with infarcts in the left cerebellum.

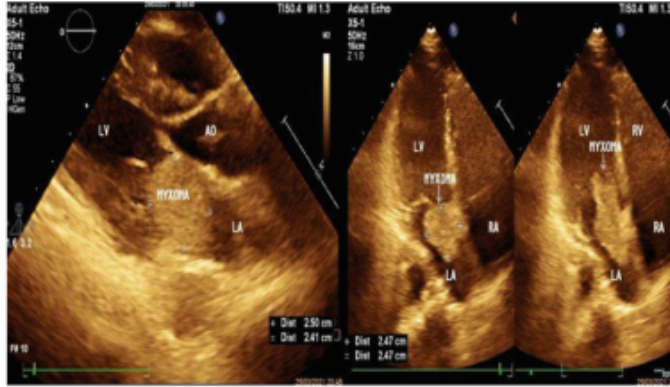


Fig. 2. Transthoracic echocardiography showed a pedunculated mass that attached to the left inter-atrial septum that moves according to the cardiac cycle with a size of 2.45 cm x 2.45 cm and suspected as a left atrial myxoma.

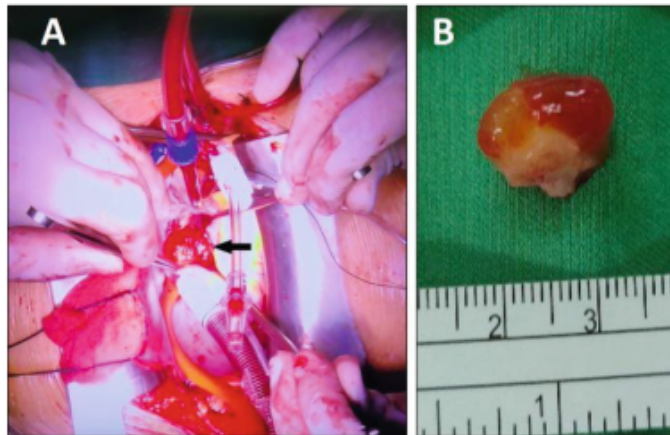


Fig. 3. A. Intraoperative image, black arrow showed myxoma mass in left atrial, B. Myxoma mass after resected.

(TTE) showed good left ventricular (ejection fraction 62,9%), mild tricuspid regurgitation, and a pedunculated mass that attached to the left inter-atrial septum that moves according to the cardiac cycle with a size of 2.45 cm x 2.45 cm and suspected as a left atrial myxoma (Fig. 2). The patient has been diagnosed with a left atrial myxoma with ischemic stroke.

Preoperative transoesophageal echocardiography (TOE) was performed and confirmed the presence of a mobile mass in the left atrial. Subsequently, surgical excision was performed via conventional median sternotomy with cardiopulmonary bypass procedures (Fig. 3). The myxoma mass, its stalk, and part of the left interatrial septal were resected, followed by the closure of the defect. Histopathological and immunohistochemical examination revealed a cardiac myxoma (Fig. 4).

Four days after the operation, the patient's condition has improved clinically and there are no postoperative complications. Followed up with echocardiography showed good left ventricular function (ejection fraction 71%) and no residual tumor mass in the left atrium (Fig. 5).

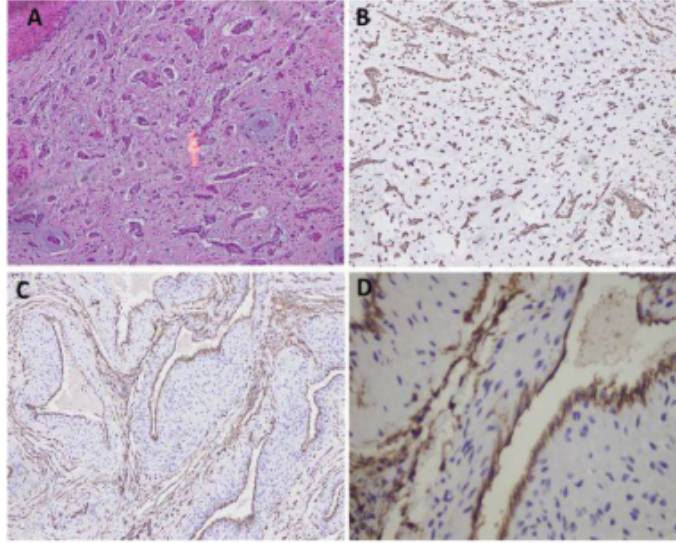


Fig. 4. Histopathological examination revealed proliferative blood vessels of varying sizes, lined with endothelial cells, some with a tombstone structure, some with thickened blood vessel walls. Some of the endothelial hyperplasias form a cord structure, the strain consists of round and oval cells, round and oval nuclei, visible nuclei, eosinophilic cytoplasm. Surrounded by a stroma of myxoid fibro collagenous connective tissue, with lightly inflammatory cells, lymphocytes, plasma, including stellate to spindle-shaped cells. Immunohistochemistry showed positive results of ERG, CD31, CD34, and Vimentin.

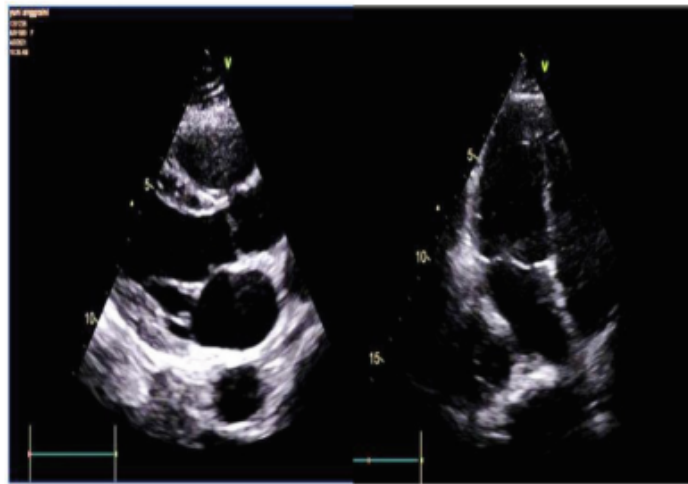


Fig. 5. Follow-up echocardiography showed good left ventricle function (ejection fraction 71%) and no residual myxoma mass in the left atrium.

Eight days after the operation, the patient was clinically improved without neurological symptoms and then allowed to go for outpatient.

3 Result and Discussion

3.1 Discussion

We reported a case of a 31-year-old woman with complaints of sudden diplopia and syncope for 4 days. She also has recurrent vertigo in the last month. Her vital sign was within the normal limit. Symptom of diplopia in patients due to ophthalmoplegia of the right eye where on neurological examination showed nerve VI, IV, and III inferior oblique paralysis. The patient also had flat right nasolabial plica (nerve VII right perifer paralysis) and tongue deviation to the right (nerve XII right central paralysis). A head CT scan showed the impression of a right-left thalamus infarct, especially the right. In this case, an ahead MRI examination was also performed which it's more sensitive than a head CT in identifying more subtle abnormalities of the brain such as stroke. A head MRI showed subacute multiple infarcts in the bilateral thalamus and bilateral centrum semiovale with infarcts with blood components in the left cerebellum.

Symptoms of syncope in patients can be due to a transient ischemic attack (TIA). Syncope is defined as a temporary loss of consciousness due to cerebral hypoperfusion, characterized by rapid onset, short duration, and spontaneous recovery [8]. One of the causes of syncope is a cardiac mass, such as atrial myxoma [8].

Symptoms of vertigo can be caused by the presence of infarct lesions in the cerebellum that often affect the PICA territory. Patients with infarcts of the PICA territory most often present with acute vertigo [9] In this case, an MRI examination of the head revealed an infarct in the left cerebellum which can cause vertigo symptoms. Atrial myxoma with cerebellar involvement is a very rare clinical presentation [9].

The diagnosis of left atrial myxoma has established a base on transthoracic echocardiography that was a pedunculated mass in the left atrium attached to the interatrial which prolapsed mass into the left ventricle in diastolic phase moment. In this case, an intermittently prolapsing mass seen echocardiography in the diastolic phase puts the patient at high risk of embolism [9] Transthoracic echocardiography is a non-invasive method that is considered as the imaging modality of choice for the diagnosis of cardiac myxoma, but the transesophageal approach provides a better picture of the location and characteristics of the tumor with a sensitivity of almost 100% [9].

The clinical manifestations of atrial myxoma are highly variable, depending on the size, location, propensity for embolization, invasion, and association with other cardiac structures [1, 2]. Patients usually present with one of the following symptoms: arrhythmias, obstruction of intracardiac flow (67%), embolic phenomena (29%), and constitutional symptoms (34%) [6, 9, 10]. However, cardiac myxomas are responsible for only 0.5% of strokes [6, 9]. Embolic manifestations occur in 20–45% of patients with cardiac myxomas. Of the 113 atrial myxoma patients with neurologic symptoms, 83% had an ischemic stroke and most often had multiple lesions (41%). Other manifestations, namely syncope (28%), psychiatric symptoms (23%), headache (15%), and seizures (12%) [6].

Lee et al. reported that 13 of 59 (22%) patients with cardiac myxoma had embolic events, of which were in the brain (18.6%), limbs (3.4%), and eyes (1.7%) [6] Embolic events occur in approximately 40–50% of patients with cardiac myxoma often to the brain due to fragments of the myxoma itself or surface emboli [9] Neurological complications

of atrial myxoma are the most common is a cerebral infarction caused by a thrombus detached from the myxoma and rarely from the tumor fragment itself [6].

In myxoma, clinical symptoms are mostly related to location, morphological characteristics, and cytokine production (especially interleukin-6). Mitral valve obstruction can cause syncope, dyspnea, and pulmonary edema followed by embolic manifestations [1, 10]. Patients may also present with nonspecific symptoms such as fatigue, cough, fever, arthralgia, myalgia, weight loss, and an erythematous rash [1, 11]. Symptoms constitutional can be mediated by interleukin-6 (IL-6) which is produced by the myxoma itself [6].

On clinical examination, left atrial myxoma can mimic many other diseases such as mitral regurgitation, pulmonary embolism, tricuspid stenosis, and tricuspid regurgitation. There were no pathogenic physical examination findings. On auscultation, an early diastolic murmur may be found as a sign of a “tumor plop” [1, 6] which is the hallmark of cardiac myxoma, but it is often indistinguishable from an opening snap [7]. In 36% of myxoma patients, no abnormalities were found on examination. Auscultation and in only 54% of cases a murmur was heard indicating mitral stenosis [9].

Myxoma produces vascular endothelial growth factor (VEGF), which contributes to inducing angiogenesis in the early stages of tumor growth [11]. Macroscopically, myxomas are yellowish, white, or brown-stemmed masses that are often covered with thrombi on examination. The surface of the myxoma consists of several fine or very fine, gelatinous villi with brittle extensions which tend to rupture spontaneously and are associated with embolic phenomena [1, 10].

Histologically, myxomas are composed of stellate, fusiform, and polygonal cells within a mucopolysaccharide stroma, [8] usually with degenerative features such as calcification and hemorrhage [1]. Calcification occurs in approximately 10% of myxoma [10] The abundance of the myxoid stroma is also associated with tumor embolization, in which the tumor ruptures and moves distally causing a blockage [1].

In general, myxoma is an emergency with life-threatening complications. A multidisciplinary approach is urgently needed for the management of cardiac myxoma patients. Once the diagnosis is established, surgery should be performed as soon as possible to prevent the risk of further tumor embolism and valve obstruction, which is very serious complication. Immediate surgical resection of the myxoma mass and its attachment to the interatrial septal wall via median sternotomy with cardiopulmonary bypass and mild hypothermia is the recommended treatment [9] The approach to resection of left atrial myxoma via median sternotomy has been demonstrated to be a safe operation, with minimal mortality and minimal recurrence rate [7] Delaying surgery will lead to embolic complications, such as limb embolism and mesenteric embolism [6].

4 Conclusion

Myxoma is the most common benign heart tumor and has varied clinical symptoms. Complications can be fatal, such as ischemic stroke, arrhythmia, heart valve obstruction, heart failure, and sudden death. A multidisciplinary approach is urgently needed for the management of cardiac myxoma patients. An echocardiological examination is very important for the diagnosis of myxoma. Cardiac surgery in the form of myxoma

resection is the definitive treatment and must be done immediately to prevent more severe complications and have a good prognosis.

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Author's Contribution. The author performed a transthoracic echocardiography examination to establish the diagnosis of left atrial myxoma, participated in joint treatment with the neurology and cardiovascular surgery department. The author collected patient data and described it in case reports with a literature review.

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