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Right Ventricular Myxoma Complicated by Stroke

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Case Report

History

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ABSTRACT

Cardiac myxoma is the most commonly diagnosed benign cardiac tumor. It is usually located in the left atrium and typically arises from the foramen ovale in approximately 75% of the cases, in the right atrium in 23%, and in the ventricles in only 2%. Symptoms depend on its size, mobility, and location. Neurological complications are seen in 20% to 25% of patients. Herein we present a case of right ventricular myxoma with patent foramen ovale which was complicated by stroke. The mass was surgically excised and patent foramen ovale was closed.

Keywords: Cardiac surgery, echocardiography, imaging, myxoma, stroke, valve disease

İnme ile Komplike Olan Sağ Ventrikül Miksoması

Olgu Sunumu

Süreç

Geliş: 02/05/2023
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ÖZET

Kardiyak miksoma kalbin en sık tanı alan iyi huylu tümörüdür. Genellikle sol atrium içinde saptanır ve olguların %75' inde tipik olarak fossa ovalisten, %23' ünde sağ atriumdan ve sadece %2' sinde ventriküllerden köken alır. Belirtiler miksomanın boyutu, hareketlilik derecesi ve yerleştiği yere bağlıdır. Nörolojik komplikasyonlar hastaların %20-25' inde gözlenir. Bu yazıda sağ ventrikül miksomaya eşlik eden patent foramen ovale saptanan ve inme ile komplike olan bir olgu sunuyoruz. Tedavide kardiyak kitle cerrahi olarak çıkarıldı ve patent foramen ovale kapatıldı.

Anahtar Kelimeler: Kalp cerrahisi, ekokardiyografi, görüntüleme, miksoma, inme, kapak hastalığı

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Introduction

Cardiac myxoma is the most commonly diagnosed benign cardiac tumor. Myxomas are usually located in the left atrium, and they typically arise from the foramen ovale in approximately 75% of the cases, in the right atrium in 23%, and in the ventricles in only 2%. Related symptoms depend on their size, mobility, and location. Neurological complications related with cardiac myxomas are seen in approximately 20% -25% of patients.[1] Herein we present a case of right ventricular myxoma with patent foramen ovale which was complicated by stroke.

Case Report

A sixty-year-old female patient presented to the outpatient clinic with exertional dyspnea and cough. Her previous medical history had no positive features. Upon physical examination, a systolic murmur was heard in the left upper parasternal border. Twelve-lead ECG revealed sinus rhythm with right axis deviation, tall R wave in V1, anterior ST segment depression and T wave negativity (Figure 1). Blood biochemistry and hemogram were normal. High-sensitivity C-reactive protein was 45.4 mg/L (N: 0-5 mg/L) and erythrocyte sedimentation rate was 25 mm/h (N: 0-30 mm/h). Transthoracic echocardiography revealed a 4.6 x 5.5 cm mass attached to the right ventricular outflow tract which protruded through the pulmonic valve during systole (Figure 2). The mass caused obstruction and CW-Doppler revealed approximately 36 mmHg systolic gradient in the right ventricular outflow tract and pulmonary artery (Figure 3). Left ventricular systolic functions were normal. There was moderate tricuspid regurgitation. In the subcostal view, a small patent foramen ovale (PFO) was detected with color Doppler (Figure 4). Transesophageal echocardiography and cardiac computerized tomography also revealed a large right ventricular mass, protruding into main pulmonary artery (Figure 5,6). There was no embolic material in pulmonary artery branches. The patient was discussed in the cardiology-cardiovascular surgery council and surgery was planned. However, the patient developed amnesia and slurred speech while waiting for the surgery day. Diffusion cranial magnetic resonance revealed a 1-1.5 cm acute ischemic lesion in the left frontotemporal area. 24-hour Holter monitoring showed no atrial fibrillation. Lower extremity venous Doppler and carotid and vertebral ultrasound were normal. Antiplatelet therapy was initiated. A preoperative coronary angiogram revealed coronary artery plaques. After the acute phase of the cerebral infarct, the cardiac mass was surgically resected and PFO was closed. The surgically excised mass was 4.5 x 5.8 cm in diameter. The macroscopic specimen of the mass demonstrated a jelly like mass including hemorrhagic areas. The microscopic examination revealed stellate cells which were spreaded in a loose myxoid stroma. There was no sign of pleomorphism, mitosis or other features suggestive of malignancy. The patient was diagnosed as right ventricular myxoma. The postoperative period was uneventful. A control echocardiogram was performed two months later, which was completely normal (Figure 7). She had no neurologic sequelae. We planned to follow the patient with annual echocardiograms as long as she has no complaints.

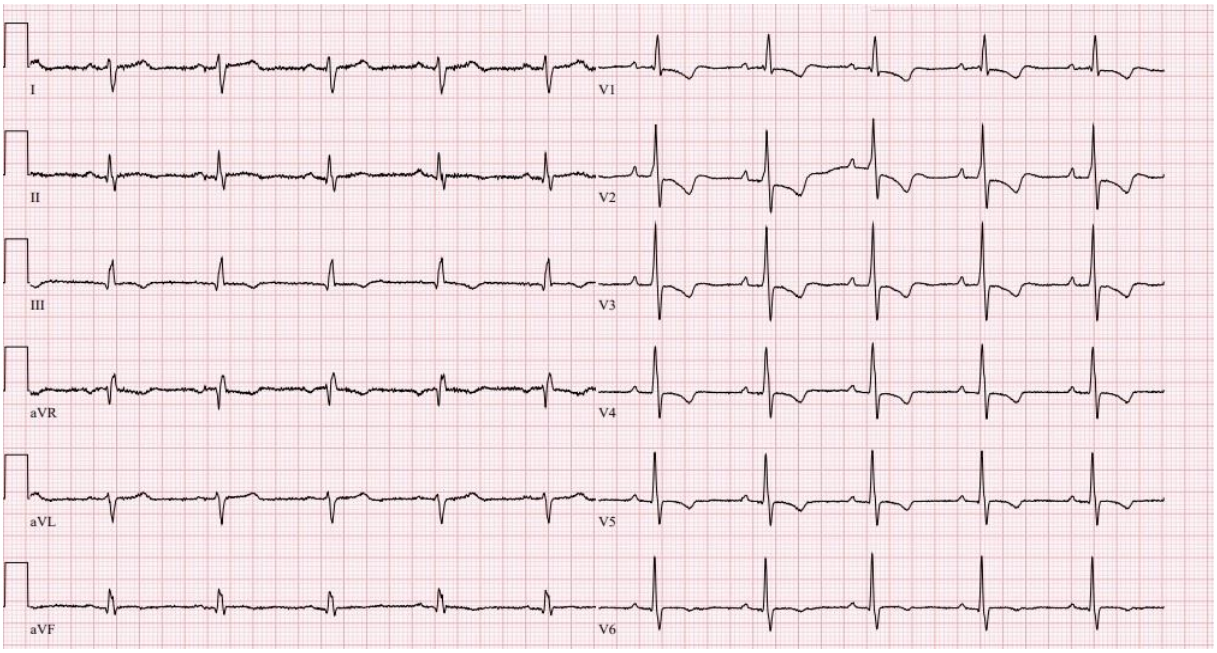

Figure 1: Twelve-lead electrocardiogram reveals sinus rhythm with right axis deviation, tall R wave in V1, anterior ST segment depression and T wave negativity.

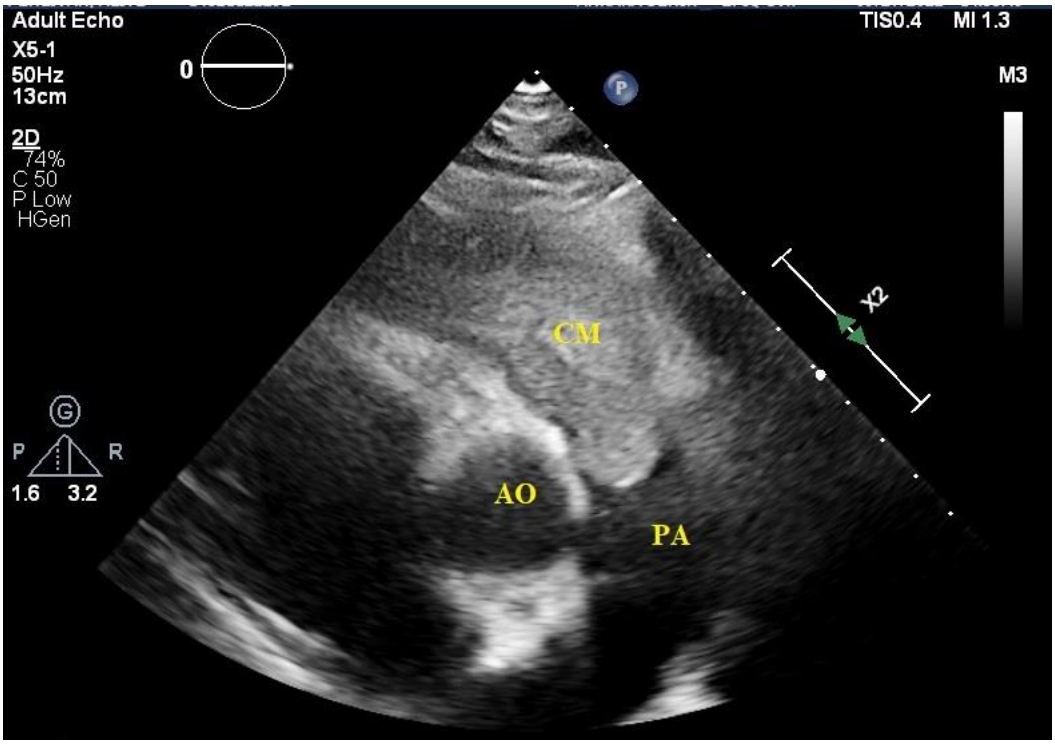

Figure 2: Transthoracic echocardiography revealing right ventricular mass protruding through the pulmonic valve during systole. Parasternal short axis view. CM: Cardiac myxoma, AO: Aortic valve, PA: Pulmonary artery.

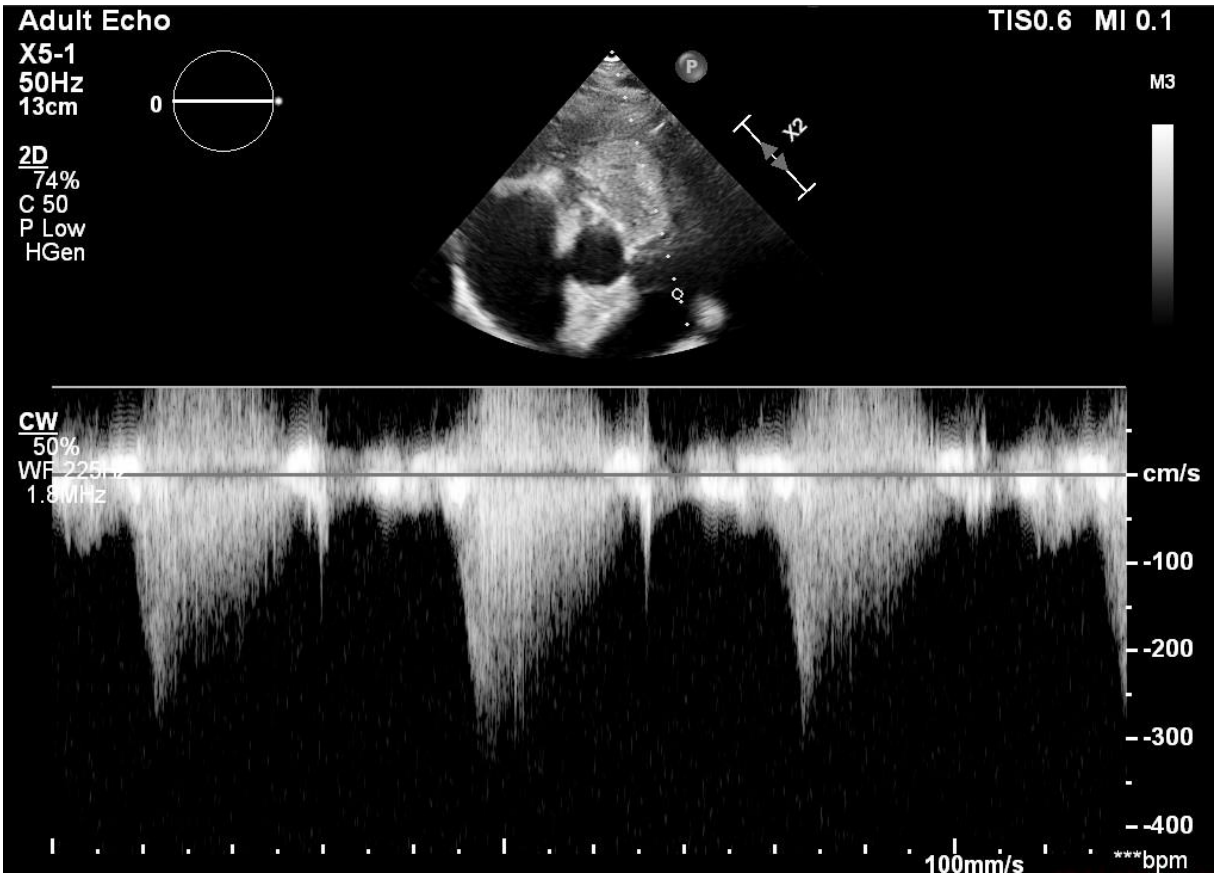

Figure 3: CW-Doppler revealing approximately 36 mmHg systolic gradient in right ventricular outflow tract and pulmonary artery. Parasternal short axis view.

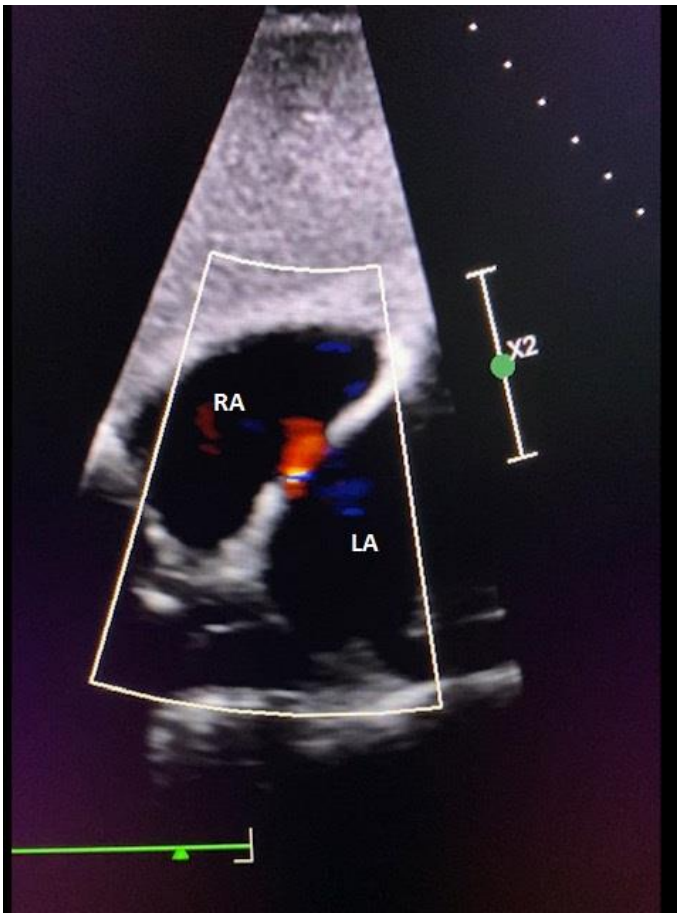

Figure 4: Transthoracic echocardiography revealing patent foramen ovale. Subcostal view. RA: Right atrium, LA: Left atrium.

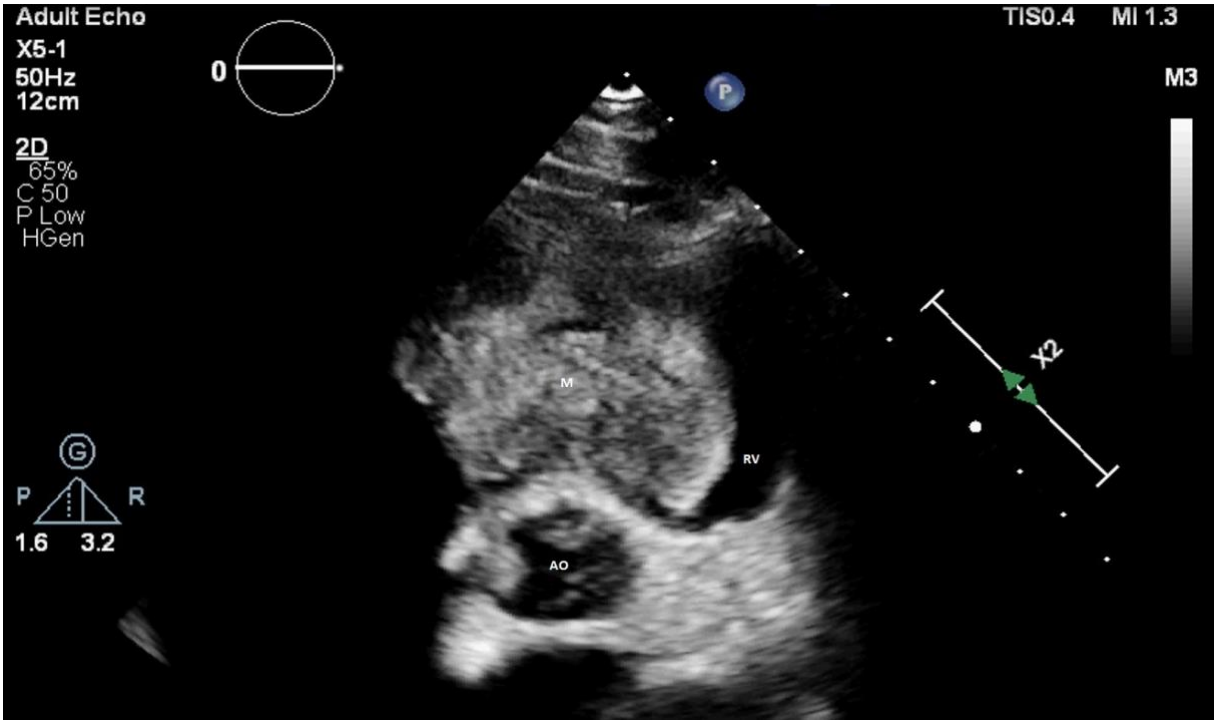

Figure 5: Transesophageal echocardiographic image of the mass. AO: Aortic valve, RV: Right ventricle, M: Myxoma

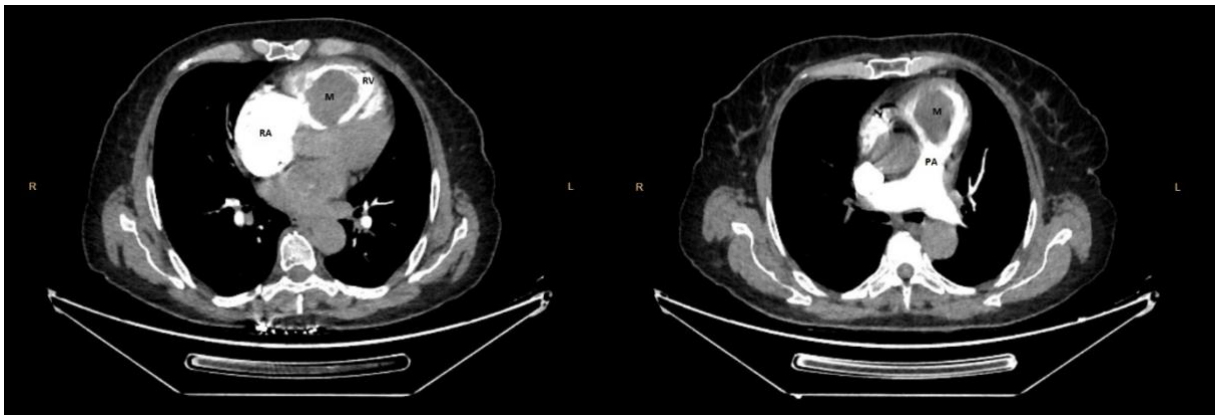

Figure 6: Cardiac computerized tomography also revealed the right ventricular mass, protruding into main pulmonary artery. PA: Pulmonary artery, RA: Right atrium, RV: Right ventricle, M: Myxoma.

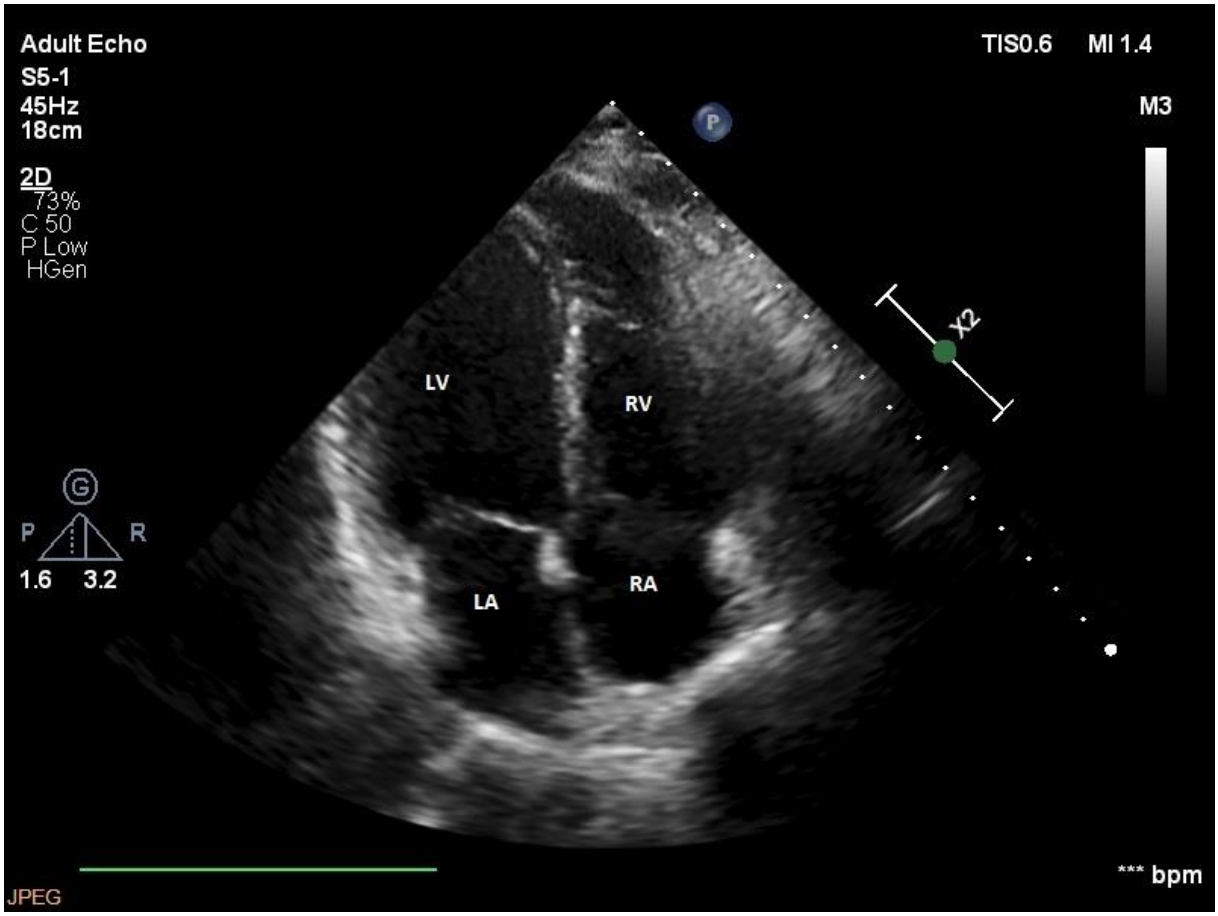

Figure 7: A control transthoracic echocardiogram was completely normal. Subcostal view. AV: Aortic valve, PA: Pulmonary artery, RA: Right atrium, RV: Right ventricle.

Discussion

We presented a case of right ventricular myxoma with patent foramen ovale complicated by stroke. The tumor was successfully resected. The clinical manifestations of ventricular myxomas can be classified as obstructive, embolic and constitutional. Clinic presentation of left-sided tumors are earlier than right-sided tumors with more severe shortness of breath.[2] Constitutional symptoms are fever, arthralgias, weight loss and Raynaud phenomenon, which may be attributed to overproduction of interleukin-6. The obstructive and embolic symptoms are different between left-sided and right-sided cardiac myxomas. For example, in patients with left-sided myxomas, obstruction may lead to pulmonary congestion, therefore dyspnea and syncope are the main complaints.

On the other hand, in patients with right-sided myxomas, peripheral edema, ascites, or superior vena cava syndrome are more common. Left-sided myxomas may cause peripheral embolism such as stroke, while right-sided myxomas can cause pulmonary embolism.[3] In a study Stefanou et al. screened 52 patients with left-sided cardiac myxoma and 13 had transient ischemic attack, ischemic stroke or retinal ischemia. Tumor friability, not the size, was associated with increased embolic risk. The embolic material can be detached tumor tissue, thrombotic material overlying the tumor, or a combination of both.[4] Our patient had elevated right heart pressures together with a PFO, and stroke probably developed due to paradoxical embolism. Patent foramen ovale, a tunnel-like structure between right and left atrium, is a common finding in general population. A right-to-left shunt and paradoxical embolism may occur when right atrial pressure increases, leading to the transit of embolic material into the systemic circulation. Our case is very rare that a right-sided myxoma caused systemic embolism through a PFO. A similar case report was written by Rao et al.[5] They presented a 62-year-old lady who experienced two episodes of acute stroke two weeks apart and she was found to have a large right ventricular myxoma and PFO. Molnar et al reported an ischemic stroke case with giant right atrial myxoma associated with PFO.[6] In another case report, a tricuspid myxoma caused paradoxical embolism through a PFO.[7] The first-line and gold standard noninvasive diagnostic modalities for cardiac myxoma are transthoracic echocardiogram and transesophageal echocardiogram. An echocardiogram enables us to evaluate preoperative localization, size, shape, mobility of the tumor as well as its hemodynamic consequences. Transesophageal echocardiography accurately detects atypical localization of myxomas. Cardiac-gated computed tomography and magnetic resonance scans provide supplementary data about the structure and function of cardiac tumors prior to surgical resection. Multimodality imaging is recommended in the diagnostic work-up.[8] The differential diagnosis for cardiac myxomas includes cardiac thrombi, fibromas, lipomas and nonmyxomatous neoplasms, the latter being malignant. About 20–25% of primary cardiac tumours are known to be malignant, the vast majority of them being sarcomas (95%), the remainder being lymphomas (5%). These malignant tumors are usually located intramyocardial and they are not pedunculated as most of the myxomas.[9]

The treatment of myxoma is surgical resection. The surgical mortality is reported as less than 5%, while the mortality rate of ventricular myxomas is slightly higher than that of atrial myxomas.[10] Once the diagnosis is made, the surgery should not be unnecessarily delayed. While awaiting to cardiac surgery, more than 8% of patients died of embolic, obstructive or other complications.[3] Our patient also had embolic stroke while awaiting to surgery, which further delayed the treatment. The need for antiaggregants and anticoagulants also complicate the perioperative period. There is no evidence for optimal time interval between cardiac myxoma related cerebrovascular event and cardiac surgery, but prolonged interval is associated with embolic recurrence.[4] Tumor recurrence may happen months or years after surgery, and its rate is approximately 5%. Recurrences usually occur in the first 4 years after surgical excision. The risk factors which are related to recurrence after surgery are; young age, family history of myxoma, inadequate surgical resection, intraoperative tumor implantation or multicentre growth.[10] There is also frequent recurrence of tumors associated with Carney complex.[11,12] Long-term follow-up with transthoracic echocardiography is recommended in all patients after tumor resection.[1]

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