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Cardiac Myxoma: A Rare Case Series of 4 Patients and a Literature Review

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Myxoma; Cardiac tumor; Cardiac mass; Imaging

Abbreviations:

CCT: Cardiac Computed Tomography; CT: Computed Tomography; ECG: Electrocardiogram; LA: Left Atrium; LV: Left Ventricle; MRI: Magnetic Resonance Imaging; RA: Right Atrium; RV: Right Ventricle; SPAP: systolic pulmonary arterial pressure; TTE: Transthoracic Echocardiography

1. Abstract

Cardiac myxomas are the most common primary cardiac tumor. Clinical findings are numerous and range from general manifestations to symptoms related to intra-cardiac obstruction or systemic embolization. Echocardiography is the key exam for the diagnosis and surgery allows tumor resection and the prevention of complications.

We present four cases of cardiac myxomas, In two of them, the left atrial myxoma was revealed by ischemic stroke. In the third case, a left atrial myxoma was incidentally diagnosed in a routine echocardiography and in the last case, an expected right atrial location of cardiac myxoma was revealed by right heart failure signs and dyspnea. All four cases were diagnosed by echocardiography and histology and all of them underwent successful cardiac myxoma resection surgery. We also reviewed clinical manifestations and elements of diagnosis and prognosis of cardiac myxomas.

2. Introduction

Cardiac myxomas are the most common benign primary cardiac tumors. About 75% are found in the left atrium, 20% in the right atrium, and 5% in the ventricles [1]. Typically, they are solitary and arise from a pedicle on or near the fossa ovalis. They occur more in females and after the third decades of life [2]. The tumor can cause intracardiac obstruction and systemic or pulmonary embolization [3,4]. The preoperative diagnosis of such tumors has been facilitated by the advent of echocardiography and is confirmed by histology. Surgical resection results improved by the advances in cardiac operations and procedures.

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3. Cases Presentation

We present four cases of cardiac myxoma.

Case 1: A 58-year-old female with history of diabetes mellitus and hypertension, presented to the emergency department with transient right hemiparesia and left facial paralysis, with a history of two similar reversible episodes three weeks before. Cerebral and thoraco abdominal Computed Tomography (CT) scan was performed revealing an acute ischemic stroke of the right middle cerebral artery and hepatic infarct. Electrocardiogram (ECG) demonstrated normal sinus rhythm with negative T-wave in anterior-septal territory. A Transthoracic Echocardiography (TTE) was performed, showing a giant hyperechoic oval mass measuring 62x33 mm. This mass was appended to the inter-atrial septum in its middle part and prolapsing in the left ventricle during diastole and almost totally obstructing blood flow. The mean pressure gradient through the mitral valve was 13 mmHg with a maximum measurement of 23 mmHg (Figure 1). Left atrium was dilated by the mass and systolic pulmonary arterial pressure (SPAP) was estimated by tricuspid regurgitation flow at 48 mmHg. The patient underwent cardiac surgery using cardiopulmonary bypass with a successful resection of the atrial mass. The postoperative period was uneventful, echocardiographic control showed a PAP normalization with no residual tumor. Histological examination confirmed the diagnosis of myxoma. The patient was discharged from the hospital on day 9 without symptoms. The 24 months echocardiographic follow up did not show any signs of recurrence.

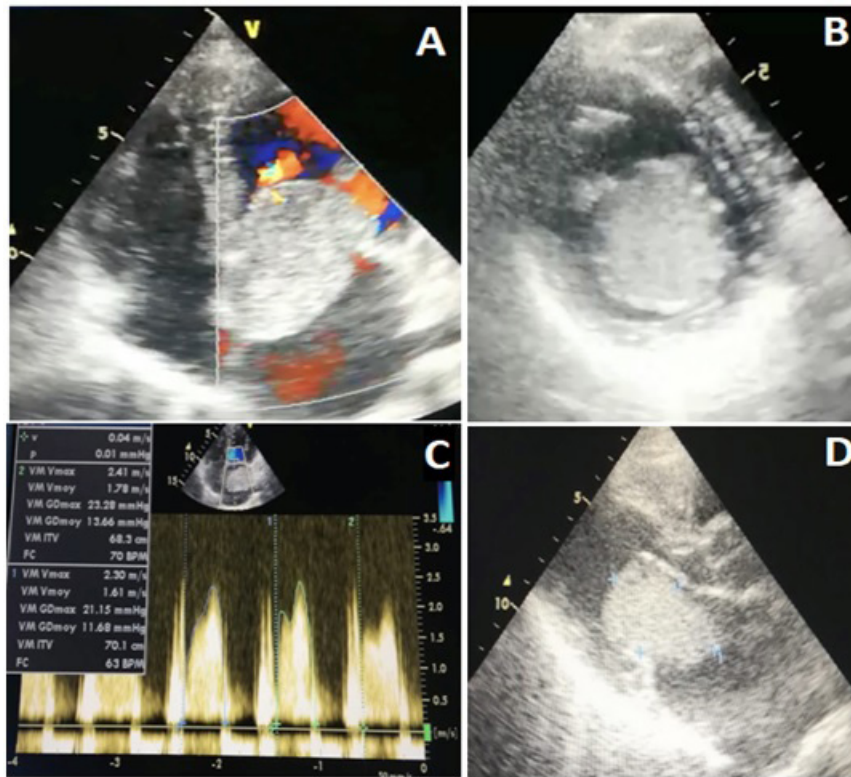


Figure 1: Transthoracic Echocardiography showing a left atrial mass attached to the inter-atrial septum. (A) Apical four chamber views showing the mass diastolic prolapsing in the left ventricle and the blood flow obstruction. (B) Basal parasternal short axis view in mitral valve level showing the giant mass. (C) Continuous-wave Doppler with mean pressure gradient through the mitral valve up to 13 mm Hg with a maximum gradient of 23 mm Hg. (D) Parasternal long axis view showing once again a left atrial mass measuring 63x32 mm.

Case 2: A 52 years old female, without any medical history, presented to the emergency department for a prolonged loss of consciousness for five hours with aphasia and abdominal pain. Immediate cerebral CT scan showed left cortico-subcortical hypodensity ranges located in fronto-parieto-occipital regions, associated with bilateral thalamic lacunar hypodensities in favor of multiple supratentorial cerebral strokes of different ages. Abdominal CT angiography revealed peripheral renal and splenic hypodense areas suggestive of infarcts. ECG demonstrated normal sinus rhythm with normal repolarization. Transthoracic echocardiography revealed a thickening of the inter-atrial septum with probable adherent thrombus measuring 5 mm. Transesophageal echocardiogram was performed for a finer study of atrial septum anatomy. It showed an echogenic oval and not very mobile mass measuring 25*15mm with a wide implantation base, attached on the left side of the atrial septum, on which a thrombus of 5 mm length is appended (Figure 2). The patient underwent surgical resection of the mass using cardiopulmonary bypass. The patient underwent a surgical resection of the mass without any incident. The anatomopathological examination revealed myxoid tumor proliferation with calretinin-expressing tumor cells, this morphological

and immunohistochemical appearance was consistent with cardiac myxoma. 6 months echocardiographic follow up didn't show any recurrence of cardiac myxoma.

Case 3: A 58 years old female with a history of diabetes and hypertension, presented to the emergency department for aggravating dyspnea associated with fever and cough evolving few days before here consultation without any embolization signs. A COVID-19 PCR test was performed given these signs and the context of COVID-19 pandemic who turned out to be positive. The patient was admitted in an intensive care unit for 15 days. A routine and systematic echocardiographic evaluation was carried out and revealed a homogeneous echogenic, elongated, mobile LA mass, measuring 65*19mm. This mass was appended to the inter-atrial septum and prolapsing into the left ventricle during diastole, without obstruction the mitral valve. Mean trans-mitral pressure gradient was 3.2 mmHg (Figure 3). The patient underwent cardiac surgery with complete resection of the mass. The histological examination showed a regular myxoid pauci-cellular proliferation. The mucoid substance is homogeneous eosinophilic, this morphological aspect corresponds to a myxoma. Post operative and 2 months echocardiographic control haven't show any residual mass.

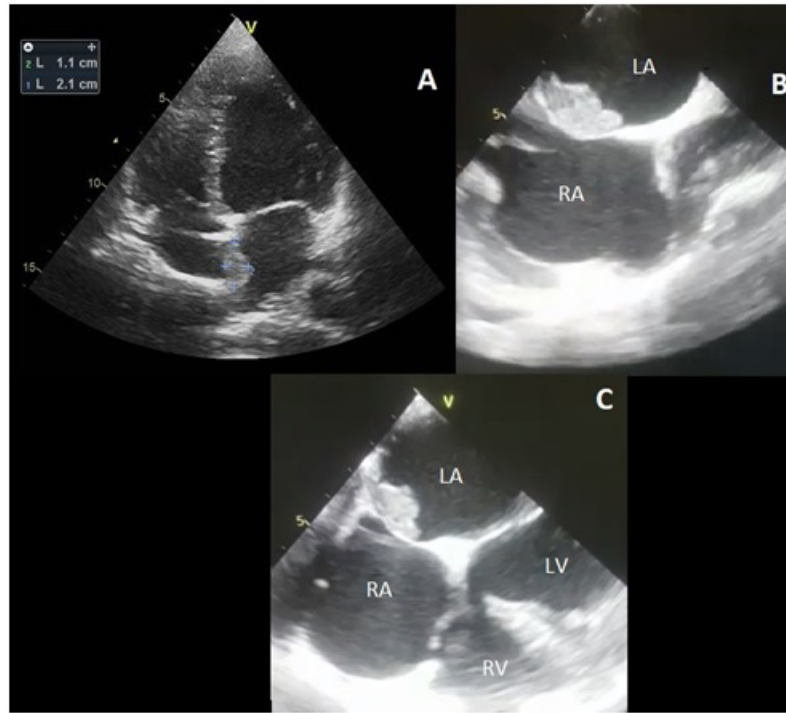


Figure 2: Echocardiography showing a left atrial mass attached to the inter-atrial septum. (A) Transthoracic Apical four chamber views showing the thickening of atrial septum. (B) Transesophageal 120° bicaval view showing the echogenic mass attached to the left side of the atrial septum with a large implantation base. (C) Transesophageal 0° four chambers view showing the mass on the left side of atrial septum with an appended small millimetric thrombus. LA: Left atrium; LV: Left ventricle; RA: Right atrium; RV: Right ventricle.

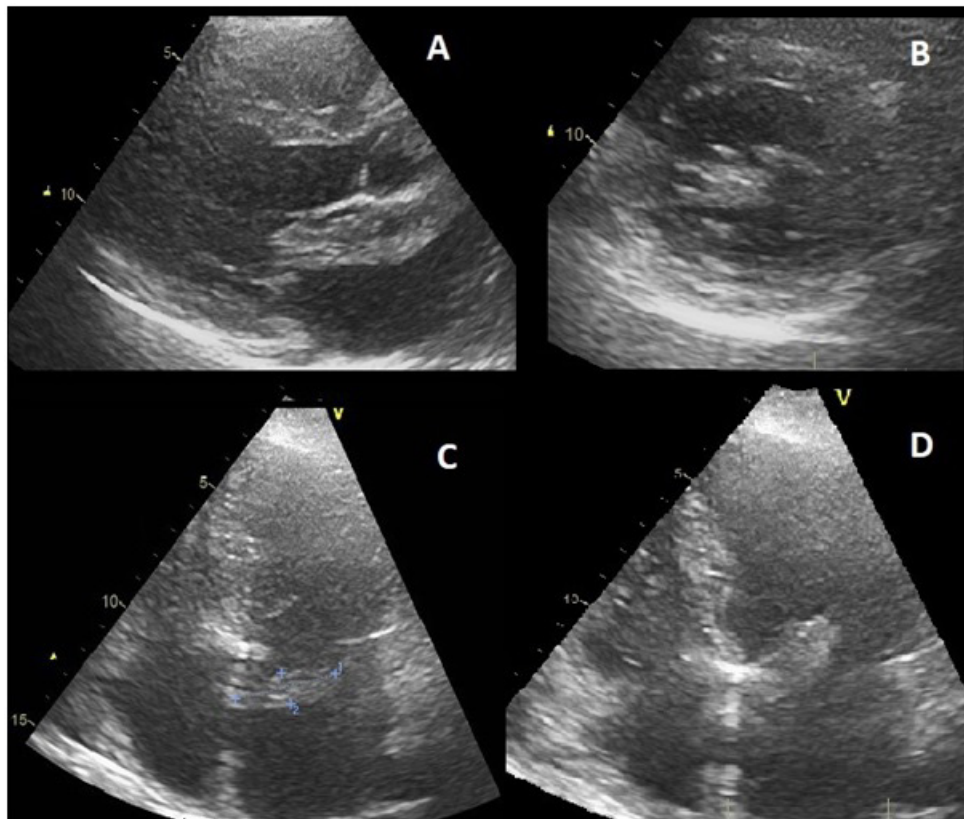


Figure 3: Transthoracic Echocardiography showing a left atrial mass attached to the inter-atrial septum. (A) Parasternal long axis view showing the mass appended to the left side of atrial septum and prolapsing in the left ventricle during diastole. (B) Parasternal short axis view showing the echogenic mass prolapsing through the mitral valve during diastole (C) Apical four chambers view showing the echogenic mass attached to the left side of the atrial septum during systole. (D) Apical four chambers view showing the echogenic mass prolapsing through the mitral valve in diastole.

Case 4: A 52 years old male with a history of diabetes, tobacco use and hypertension, presented for dyspnea on exertion since few months. Physical examination revealed right heart failure signs with bilateral lower limbs edema, basal right pleural effusion syndrome, pericardial friction rub. The ECG showed a diffuse micro-voltage. chest X-ray revealed a moderate right pleural effusion. Transthoracic echocardiography showed a huge echogenic mass filling the entire RA measuring 80*62mm. This mass appeared to be appended to the atrial septum and was obstructing the tricuspid valve during diastole. The mean pressure gradient through the tricuspid valve was 12 mmHg. There was a pericardial effusion of

medium abundance towards the right ventricle (Figure 4). There was no evidence for pulmonary embolism on thoracic computed tomography (CT) angiography. Tumor markers were all negative and the Body CT didn't show any primary or secondary tumor locations other than this mass. Cardiac surgery was carried out after a normal coronarography, the mass appeared attached to the right side of atrial septum during intraoperative assessment. Anatomopathological study showed a myxoid tumor proliferation. Dyspnea and right heart failure signs regressed after cardiac resection surgery with no recurrence on echocardiographic control at one year.

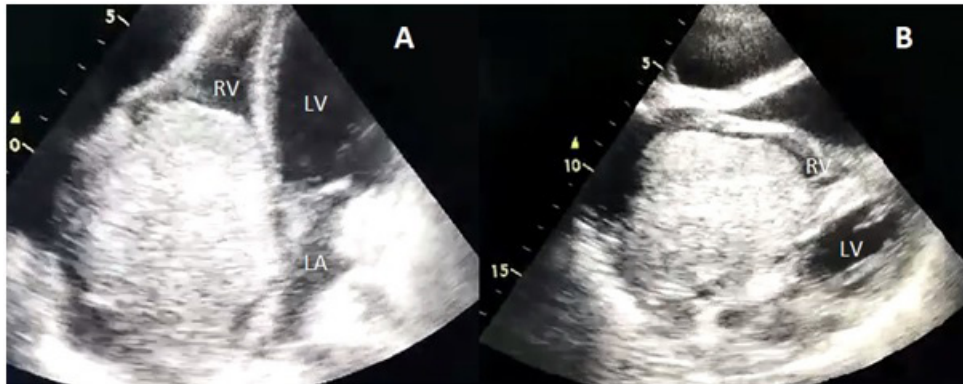


Figure 4: Transthoracic echocardiography showing a right atrial mass was obstructing the tricuspid valve during diastole (A) Apical four chamber views a giant mass of right atrium, (B) Subcostal view showing the giant mass of the right atrium with moderate pericardial and pleural effusion. LA: Left atrium; LV: Left ventricle; RV: Right ventricle.

4. Discussion

Cardiac myxoma is the most common 'benign' cardiac tumor and may have varying clinical presentations. The classic triad of clinical presentation includes: obstructive cardiac symptoms; embolic signs and symptoms; and constitutional or systemic manifestations. Although the identification of cardiac masses is usually made using imaging, the final diagnosis of cardiac myxoma is confirmed following surgical resection of the cardiac tumor and histopathological assessment of the sample [5].

Cardiac myxoma is rare in children and is less frequent in blacks [6]. It can occur at any age but are most common age interval is between 30 and 60 years, with a female predominance [7]. The mean age of our four patients was 55 years at the time of surgery and three of our patients were female which agrees with data from the literature.

Cardiac myxoma is located in 75% of cases in the left atrium (LA), 23% in the right atrium (RA), and only 2% in the ventricles. The most common site of attachment is the fossa ovalis. Multiple locations are not very common, but can be present in 50% of familial forms [8]. In our patients, three cases had a left atrial location and one case involved a right atrial myxoma.

Clinical manifestations of cardiac myxoma are numerous, they depend on the location, shape, size, growth rate and friability of the tumor. They can be related to intra-cardiac obstruction includ-

ing signs of dyspnea, orthopnea, malaise, palpitations or syncope, or they can be may be secondary to systemic emboli [9] which are either tumoral when the myxoma is friable, or thromboembolic formed on the tumor surface. Embolic locations are mostly cerebral, revealed by ischemic stroke or retinal artery embolism responsible for sudden drop in visual acuity [10, 7]. Emboli can also affect the upper and lower extremities as well as the coronary arteries and cause myocardial infarction. In addition cardiac myxoma can generate general manifestations including fever, anemia, fatigue, joint pain, weight loss, and even cachexia and other systemic reactions [11]. Cardiac myxoma can also be discovered incidentally on cardiac or thoracic explorations carried out for another purpose. In our patients, two cases were revealed by ischemic stroke, one case by signs of heart failure and one case was discovered incidentally during routine transthoracic echocardiography during an episode of COVID19 infection.

Transthoracic two-dimensional echocardiography gives the initial clue toward the presence of a cardiac mass, its location, echogenicity, morphology, measurements, mobility, its potential for embolization and the presence of a possible valvular obstruction. However, in some cases the diagnosis can be difficult, especially in patients with poor echogenicity. The delineation of left atrial tumor insertion site and tissue characteristics may be limited field using transthoracic echocardiography. Trans-esophageal echocardiography provides further details about the location as well as mobility,

it offers a considerably better delineation of the tumor insertion site (commonly in the left atrial septum) and tissue characteristics especially for small-sized lesions [12]. It should be considered in any complicated case of cardiac mass [13]. Trans-esophageal echocardiography may also improve the detection of other major cardioembolic sources (e.g., intra-cardiac thrombus, vegetations or aortic arch plaque), as well as less common potential sources (e.g., patent foramen ovale, atrial septal aneurysm or left ventricular aneurysm) [14] and It can also show multiple cardiac locations of the tumor. In our second patient, transthoracic echocardiography showed a thickening of atrial septum, Trans-esophageal echocardiography performed for suspicion of a cardiac mass to better study the atrial septum and show the insertion of the mass on its left side.

In recent years Cardiac Computed Tomography (CCT) is increasingly utilized for evaluation of cardiac masses. It helps confirming the clinical and/or echographic suspicion of myxoma and determining the location and the size of the tumor. It can also be used to distinguish left atrial myxomas from left atrial thrombi. When an associated intra-cardiac thrombus is detected, anticoagulant therapy must be started while waiting for the surgery, however it can only have an impact on thromboembolic migration and does not prevent tumor emboli [15]. Additionally, myxoma may be incidentally diagnosed on CCT when the test is performed for other reasons like assessment of coronary artery disease. The advantages of CCT include rapid acquisition, high and isotropic spatial resolution and the ability to assess coronary anatomy which may be required prior to cardiac surgery. Another important benefit of CCT in myxoma patients is comprehensive information about cardiac anatomy, which can be paramount for cardiothoracic surgical planning [16]. Cardiac magnetic resonance imaging (MRI), by improving tissue characterization, is likely to add key information before surgical excision of large cardiac masses detected by echocardiography [17].

After diagnosis, surgery should be performed urgently, in order to prevent complications such as embolic events or obstruction of the mitral orifice. Surgical excision of cardiac myxoma carries generally a low operative risk and gives excellent short-term and long-term results [18].

Follow-up examination, including echocardiography, should be performed regularly due to risk of recurrence. The estimated recurrence rate of sporadic myxoma after cardiac resection surgery is 2 to 3%. Recurrences usually appear during the first four years, although they can emerge within a few months to several years after surgical excision. Unlike primary myxomas, which are more frequent in women, tumor recurrence appears to be more frequent in men [19]. In our four patients we had a follow-up between 6 months and 24 months, no recurrence was detected over this period.

5. Conclusion

Cardiac myxomas are the most common primary cardiac benign tumors, that may cause a wide range of clinical symptoms not only such as cardiac disease, but also general manifestations. It is most frequently found in the left atrium, but others cardiac locations can be observed. Diagnosis is rarely made only on clinical findings, because there are no specific physical signs or symptoms. Two-dimensional transthoracic and trans-esophageal echocardiography, are the technique of choice for diagnosis and follow-up of this tumor. Surgery generally has good results and the anatomopathological study confirms the diagnosis of cardiac myxoma. Recurrence is possible, which makes periodic echocardiographic monitoring necessary after tumor resection.

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